MISSOURI BIRTH DEFECTS 1996 – 2000

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Introduction

This report presents data on birth defects and other selected conditions among 1996-2000 Missouri resident live births, obtained from the Missouri birth defects registry. Birth defects, defined as abnormalities of structure, function, or biochemistry originating before birth, are a major but understudied contributor to infant mortality and morbidity. Approximately one in four Missouri infant deaths have a birth defect listed as cause of death. The true proportion is likely to be greater, as less than half of infant deaths are autopsied, and birth defects may be contributing factors in infant deaths with other listed causes.

While the genetic basis of some defects is known, and a few teratogens have been identified, the cause of most birth defects remains unknown and most cannot, at this time, be prevented. A notable exception is the ability to prevent 50-70 percent of neural tube defects (NTDs) with adequate folic acid intake before and during early pregnancy. Recent research indicates that increased folic acid intake may also reduce the risk for other birth defects, including oral clefts and congenital heart and urinary tract defects. For further discussion of folic acid and NTDs, see pages 9-10).

The Missouri Department of Health and Senior Services (DHSS) established the birth defects registry to monitor the incidence of birth defects among Missouri infants, to provide data for epidemiological studies, surveillance for potential effects of environmental contamination, and to support and evaluate state programs serving ill and disabled children. Data are available for births since 1980, but improvements in the data set make data for births since 1993 much more complete than data for earlier years (Appendix A). In 1998, DHSS was awarded a cooperative agreement with the Centers for Disease Control and Prevention (CDC), which enabled DHSS to improve the timeliness of its data and to use registry data to improve access to state-provided services.

Data presented in this report are for liveborn Missouri residents. Defects must be diagnosed in the first year of life in order to be included in the registry. The registry is a data collection effort in which data available from a number of sources are merged. Data sources currently available are birth certificates, infant death certificates, newborn patient abstracts, pediatric patient inpatient and outpatient abstracts, the DHSS data base for children enrolled in programs such as Children with Special Health Care Needs, and the Department of Mental Health First Steps (early intervention) data base. Defects reported in each component are linked to the birth certificate to avoid duplication. The birth certificate also provides important demographic data not available in most of the individual components and provides a basis for producing rates and comparing attributes of infants with and without birth defects.

The Missouri birth defects registry is a passive data collection system; case finding is limited to reports provided to DHSS from the sources listed above, and verification of reported defects is not undertaken. It is probable that some Missouri infants have birth defects diagnosed in the first year of life that are not reported on any of the data components and are not, therefore, included in the registry; these are termed false-negative cases. Conversely, the registry will include cases termed false positive: spurious reports of birth defects that are the result of mis-diagnosis, mis-

coding, or tentative diagnoses that are later discounted. Some birth defects are not diagnosed in infancy and are therefore not included in the registry. Anomalies also affect pregnancies not resulting in live births. Birth defects often result in intrauterine death. The registry includes limited data on defects among spontaneous fetal deaths of 20 or more weeks gestation, but those data are not included in this report. Additionally, prenatally diagnosed defects often result in an elective termination of pregnancy. No registry data are available for elective terminations. Please read Appendix A for a further description of the birth defects registry and its components.

The purpose of this report is to provide descriptive statistics on the prevalence of birth defects in Missouri, by characteristics such as sex, race, maternal age, and area of residence; the association of birth defects with low birth weight, preterm delivery, and infant death; and participation of infants with defects in the Medicaid, Children with Special Health Care Needs, and First Steps programs.

Because of the complexities of the data, the reader is advised to read the technical notes and appendices in order to properly understand and use the statistics presented in this report.

Report Methodology and Terminology

The study population is 1996-2000 liveborn infants whose mothers resided in Missouri at delivery. It includes infants of Missouri residents born in other states.

Diagnostic Definitions

Defects are coded according to the International Classification of Disease-9th Revision (ICD-9). The ICD-9 codes comprising each diagnostic category presented in this report are listed in Appendix B. There is no standard definition of the specific conditions that should be included in a comprehensive, or overall, birth defect category. For this report, overall birth defect data refer to the number or rate of infants with one or more conditions classifiable to the ICD-9 congenital anomalies section (codes 740-759), excluding some insignificant anomalies listed in Appendix B.

Several other conditions not included in comprehensive birth defect statistics are listed at the bottom of tables detailing defects by type (Tables 2-6), under the heading "Other Selected Conditions." These conditions are primarily of genetic or prenatal origin, but also include disorders that may arise postnatally, e.g., cerebral palsy and intracranial hemorrhage.

Rates, confidence intervals, and statistical significance

Unless otherwise specified, rates are per 10,000 live births. These statistics are sometimes based on a small number of cases and should be used cautiously. A rate is an estimate of the prevalence of a given condition. Exact binomial confidence intervals for overall birth defect rates by year of birth, race, sex, and maternal age are listed in Table 1. The 95 percent confidence interval represents a range of values within which we can state, with 95 percent confidence, the true rate lies. When the confidence intervals for two rates do not overlap, the observed difference in rates is considered statistically significant. Tables 2-4 and 7-9 include notations of statistically significant differences in rates, but not confidence intervals.

Methodology for determining statistical significance is that described above, i.e., calculation and comparison of 95 percent exact binomial confidence intervals.⁵

Race

Race-specific data are based on maternal race as listed on the birth certificate. Table 1 and Figure 2 include overall birth defect statistics for three race categories: white, black, and other. Defect-specific data for white and black infants are provided in Table 3; detailed data for other races are not provided because of the small number of defects for other race categories.

Maternal Age

Maternal age is that of the mother's age at delivery. Defect-specific data by age are presented in Table 4.

Low birth weight, preterm births, infant death

Data on two prematurity indicators are provided: low birth weight (LBW, less than 2,500 grams) and preterm (less than 37 weeks gestation). Of the two indicators, LBW is generally considered the more reliably reported, but because birth defects may affect birth weight independently of prematurity, both indicators are provided in this report. Infant deaths are those after birth and before the first birthday. Statistics are expressed as the number and percent of all LBW, preterm births and infant deaths that have one or more birth defect (Figure 5), or the number and percent of births with a defect that are LBW, preterm, and infant death (Table 5).

Medicaid

Medicaid statistics (Table 6) include the number and percent of births with defects to mothers who were Medicaid recipients during pregnancy, as listed on the birth certificate. Most infants of mothers enrolled in Medicaid are also enrolled at birth, and others not enrolled at birth may become Medicaid recipients later.

Children with Special Health Care Needs (CSHCN)

This DHSS program reimburses health care providers for many of the diagnostic and treatment services required for children with conditions including, but not limited to, many of the defects presented in this report. Eligibility is based on family income. While the CSHCN program provides assistance to children from birth to age 21, the statistics presented in this report (Table 6) are limited to the number and percent of infants enrolled in the first year of life.

First Steps

First Steps is an early intervention program for children, birth to age 3, who have delayed development or diagnosed conditions that are associated with developmental disabilities. First Steps is a Department of Elementary and Secondary Education program, and was developed in response to Part C of the Individuals with Disabilities Act. Among services provided are evaluation, counseling, and special instruction and therapy. There is no income requirement for participation in the program. For the years presented in this report, DHSS and the Department of Mental Health provided First Steps services and enrolled children in the program. Although the program offers services from birth to age three, statistics provided in this report (Table 6) are the number and percent of infants enrolled in the program before the first birthday.

County of residence

County of residence refers to the mother's residence at delivery. The number, rate, and statistical significance of the variation from the state are provided for four birth defect categories: overall birth defects (Table 7); neural tube defects (Table 8); oral clefts (Table 9); and Down syndrome (Table 10). Quartile rankings by county for these four categories are also displayed in Maps 1-4. If fewer than 80 percent of a county's births were linked to a newborn patient abstract, data are flagged and noted to be unreliable, and the county's rate is not included in quartile rankings. In Table 11, the frequency of selected birth defects is listed for each county.

County-specific data should be used with special caution. In addition to the problem of rates based on small numbers of events, other factors influence rates on a county level. Patient abstract data summarizing diagnoses made at birth and during subsequent hospitalizations are the most important source of birth defect data; some counties have a significant number of births in out-of-state or military hospitals for which patient abstract data were unavailable (see Appendix C). Additionally, differences in rates may only represent diagnostic coding practices prevalent in certain hospitals.

Data Highlights

Year of Birth

Overall birth defect rates ranged from 549 to 575 per 10,000 live births over the five-year period presented in this report (Figure 1). Of interest is the generally downward trend for neural tube defect rates from 8.1 per 10,000 in 1996 to 6.0 in 2000, with the lowest rate of 5.3 recorded in 1999 (Table 2). For a further discussion, see pages 9-10.

Figure 1 Birth Defect Rates by Year of Birth Missouri Resident Data 1993-2000

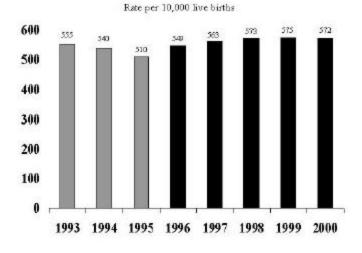
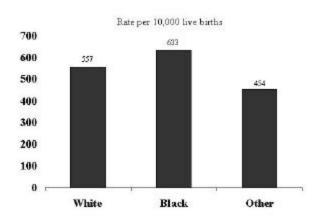


Figure 2 Birth Defect Rates by Race Missouri Resident Data 1996-2000



Race

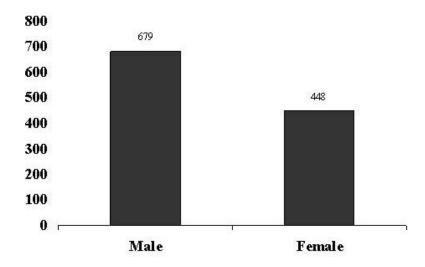
The overall 1996-2000 birth defect rate is significantly higher for black infants than for white infants (Figure 2). The rate for other races was significantly lower than both the white and black rates; 70 percent of births in the other category were to Asian or Pacific Islander women. Among defects reported significantly more frequently for black infants were hydrocephalus, microcephalus, anomalies of eye, ear, face, and neck, heart and other circulatory anomalies, and integument anomalies (Table 3). White infants had significantly higher rates of oral clefts, digestive system anomalies, hip dislocations, and skull and facial bone anomalies. Among listed conditions not included in the overall birth defect statistics, congenital syphilis, sickle cell anemia, cerebral palsy, epilepsy, and intracranial hemorrhage were reported significantly more frequently for black infants.

Sex

For 1996-2000, male infants had a 52 percent higher overall birth defect rate than females (679 v. 448, respectively, Figure 3). Most of the difference in rates reflects higher rates of genital and urinary organ defects among male infants (Table 3), but male infants also had significantly higher rates for a number of other defect categories, including hydrocephalus, heart and other circulatory/respiratory anomalies, cleft lip with or without cleft palate, pyloric stenosis, clubfoot, and skull and facial bone anomalies. Female infants had higher rates of microcephalus and congenital hip dislocation.

Figure 3
Birth Defect Rates by Sex
Missouri Resident Data 1996-2000

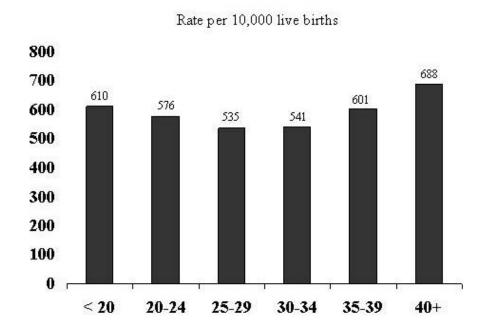
Rate per 10,000 live births



Maternal age

The overall risk of birth defects is increased at both ends of the maternal age spectrum (Table 4, Figure 4), highest for infants of mothers aged 40 plus, but also elevated for mothers less than 25. Chromosomal anomalies account for most of the increased risk for women aged 35 plus, but infants of older mothers also had significantly higher rates of heart and other circulatory/respiratory defects, atresia or stenosis of the small intestine, and urinary organ anomalies. Compared to infants of mothers aged 25-34 years, infants of both younger and older mothers were at higher risk of central nervous system defects. Infants of women less than 25 also had higher rates of atrial septal defects, digestive system, and musculoskeletal and integument defects. Additionally, they were at higher risk for congenital pneumonia and intracranial hemorrhage.

Figure 4
Birth Defect Rates by Maternal Age
Missouri Resident Data 1996-2000



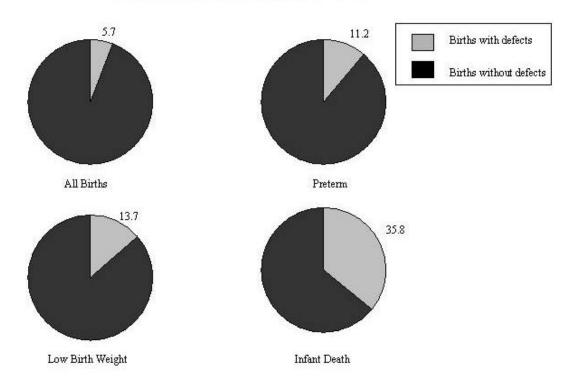
Low Birth Weight and Preterm Births, Infant Death

As shown in Figure 5, while births with defects represented only 5.7 percent of all births, they represented 11.2 percent of preterm births, 13.7 percent of low birth weight (LBW) and 35.8 percent of infant deaths (including infants with defects whose listed cause was not a birth defect).

The number and percent of births with defects that are LBW, preterm, and infant deaths are listed in Table 5. Overall, births with defects had an 18.7 percent LBW rate and a 20.1 percent preterm rate, in comparison with LBW and preterm rates of 6.7 and 9.1 percent, respectively, for infants without defects. Most defect categories had elevated rates for both indicators. In general, LBW and preterm rates are also high among infants with the other conditions presented but not included in birth defect statistics, particularly intracranial hemorrhage, because prematurity is a major risk factor for development of this condition.

In addition to an encephaly, an invariably fatal condition, defects with infant death rates in excess of 60 percent are hypoplastic left heart syndrome, agenesis of the lung, and trisomies 13 and 18. Overall, 4.8 percent of births with defects died in infancy, compared to 0.5 percent of infants without birth defects noted.

Figure 5
Percent of Total Births, Preterm and Low-Birth Weight Births, and
Infant Deaths with One or More Birth Defects
Missouri Resident Data 1996-2000



Medicaid, CSCHN and First Steps Participation

The number and percent of births participating in the Medicaid, Children with Special Health Care Needs (CSHCN) and First Steps programs are listed in Table 6. The 44.1 percent rate of Medicaid participation at birth among all infants with defects is slightly higher than the 40.5 percent rate for all births. Among infants with gastroschisis and other abdominal wall defects, 63.7 percent were Medicaid participants, reflecting the preponderance of gastroschisis among infants of poor and young mothers.

In the first year of life, 5.1 percent of infants with defects were enrolled in the CSHCN program, including 25.9 percent of infants with spina bifida and 22.9 percent of those with hydrocephalus. The program also served 23.2 percent of infants with cerebral palsy.

The First Steps program enrolled 11.0 percent of infants with defects in the first year of life. These include 57.6 percent of infants with spina bifida, 54.3 percent of those with hydrocephalus, and 68.6 percent of Down syndrome infants. In addition, 83.2 percent of infants with infantile cerebral palsy and 60.8 percent of infants with epilepsy were enrolled as infants in the First Steps program.

County of Residence

Overall birth defect rates by county of residence are listed in Table 7, and pictured in Map 1. Counties with an overall birth defect rate in the highest quartile and significantly higher than the rest of Missouri were Adair, Camden, Cape Girardeau, Christian, Cole, Dent, Phelps, and Stone. Although the overall birth defect rates for Franklin and Greene counties were significantly high in comparison with the state rate, the rates for these counties were not in the top 25 percent of overall birth defect rates by county. Caution should be used when evaluating differences in rates between counties, especially the overall birth defect rate. This category includes many diagnoses, including relatively minor defects, and differences between counties may largely reflect differences in reporting practices among the hospitals utilized by a county's patients.

Neural tube defects are distributed relatively evenly throughout the state (Table 8, Map 2). Only Osage County's NTD rate was significantly higher than the state rate. The counties making up the major metropolitan (St. Louis and Kansas City) areas have a somewhat lower NTD rate than the rest of Missouri (6.1 v. 6.8, respectively), but the difference is not statistically significant.

Oral cleft rates are significantly higher in the rural counties (Table 9, Map 3). Oral clefts are somewhat more common in white than black infants (Table 3), and rural Missouri counties have a much higher proportion of white infants. However, the racial make-up of the two groups only explains a portion of the difference in oral cleft rates between metropolitan and non-metropolitan counties. The rates for both white and nonwhite infants in nonmetropolitan counties are higher than the corresponding metropolitan rates. Knox, Scott, and Wayne counties had significantly elevated oral cleft rates.

Down syndrome rates by county are shown in Table 10 and Map 4. No county had a significantly elevated rate of Down syndrome. Metropolitan and non-metropolitan rates are similar.

Neural Tube Defects and Folic Acid

The neural tube is an early embryonic structure that develops into the brain, spinal cord, and supporting bone structures. Neural tube defects (NTDs) occur when the neural tube fails to develop properly. These defects occur very early in pregnancy, between the 17th and 30th days after conception, that is, before most women know that they are pregnant.

The two major NTDs are anencephaly and spina bifida. Anencephaly is a fatal condition in which the brain is absent or fails to develop completely. Anencephalic pregnancies often result in fetal death, and liveborn anencephalic infants die soon after birth.

Spina bifida occurs when the lower end of the neural tube fails to close, resulting in improper development of the spinal cord and backbones. A sac containing spinal fluid, and in some cases a portion of the spinal cord, may protrude from the back. Most babies born with spina bifida survive infancy; the 1996-2000 Missouri survival rate was 93 percent. Surviving infants face varying degrees of morbidity and disability, including paralysis of the legs, hydrocephalus, bladder and bowel control problems, and learning disabilities.

In 1992 the Center for Disease Control and Prevention (CDC) concluded that 50-70 percent of NTDs could be prevented by daily consumption of 400 mcg (0.4 mg) of folic acid before and during early pregnancy. Folate is one of the B vitamins and occurs naturally in many foods, including eggs, beans and peas, oranges, and many green vegetables, but it is difficult for women to consume the recommended amount through diet alone. Most multi-vitamins contain the recommended amount of folic acid. The Food and Drug Administration authorized optional folic acid fortification of enriched grain products beginning March 1996 and mandatory fortification beginning January 1998. Fortification was expected to add 100 mcg of folic acid to the average daily diet.⁷⁻⁸

NTDs in Missouri

Each year in Missouri, about 10 infants are born with anencephaly and 38 with spina bifida. The number of affected pregnancies is considerably higher. Each year NTDs are noted on approximately 8 fetal death certificates (completed for pregnancies of 20 or more weeks gestation). Other NTD-affected pregnancies result in earlier fetal loss for which no data are available. Many NTD cases are detected prenatally and are electively terminated. Although the DHSS birth defects registry does not include data for elective terminations, we were able to obtain the number of 1997-99 NTD-affected elective terminations from five major abortion providers. For those years, the facilities collectively averaged 9 terminations per year of NTD-affected pregnancies.

The rates of spina bifida among 1993-2000 Missouri live births are displayed in Figure 6, and those for anencephaly in Figure 7. The 1997-2000 spina bifida rate of 4.6 per 10,000 is 32 percent lower than the 6.8 per 10,000 rate for 1993-96. The decline in the spina bifida rate in Missouri is temporally associated with folic acid grain fortification, although other factors may be associated with the decrease. Anencephalus did not have a similar decline. Between 1993 and 2000, the rates for anencephaly ranged from 0.7 to 2.9 per 10,000, with no temporal trend.

Similar trends in NTDs have been observed elsewhere. Birth certificate reports of NTDs in 45 states revealed that from 1995 to 1999, there was a statistically significant 23 percent decline in reported cases of spina bifida, with an 11 percent decline in anencephalus that was not statistically significant.⁹

Figure 6 Spina Bifida Rates per 10,000 Live Births Missouri Resident Data 1993-2000

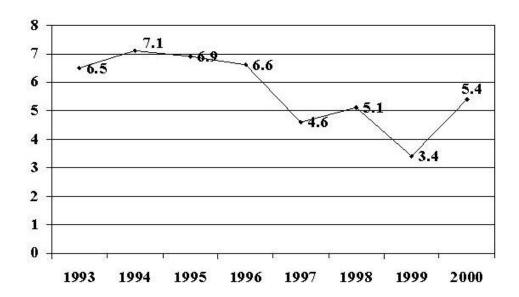
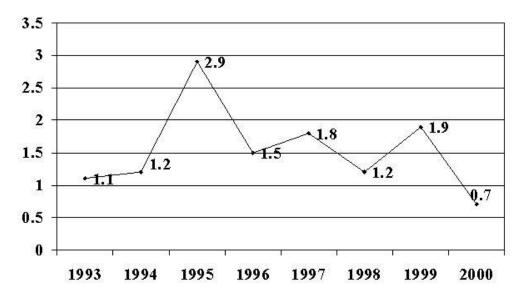


Figure 7 Anencephaly Rates per 10,000 Live Births Missouri Resident Data 1993-2000



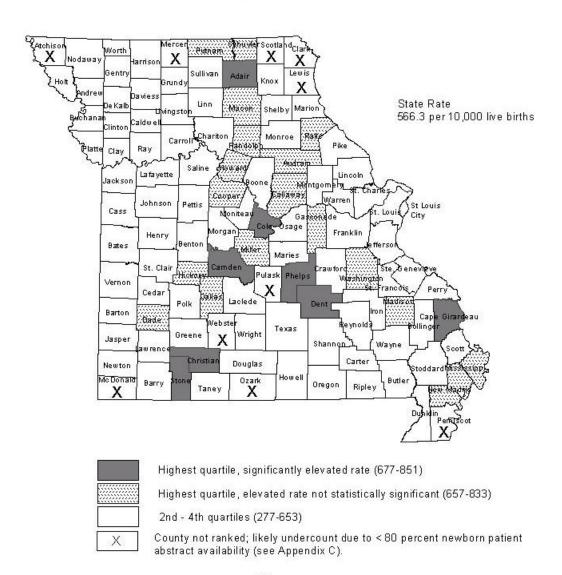
Recommendation

Neural tube defects are among the most serious of birth defects, and the identification of the effectiveness of adequate folic acid intake in preventing NTDs is one of the most promising public health developments in recent years. Significant declines in the birth prevalence rate of spina bifida, but not anencephalus, have been observed in Missouri and other states, and may be associated with folic acid fortification of grain products. However, the level of folic acid fortification of grain products does not result in the consumption by most women of the recommended daily intake of folic acid. The U.S. Public Health Service strongly recommends that all women capable of becoming pregnant—not just those planning a pregnancy—should consume 400 mcg (0.4 mg) of folic acid daily.⁶ A multi-vitamin containing folic acid is an effective method of obtaining the recommended daily intake. Folic acid supplementation is urged for all women capable of childbearing because NTDs occur too early in pregnancy to prevent after a pregnancy is recognized, and because half of all pregnancies are unplanned. Future research may establish the effectiveness of folic acid in reducing the risk of other types of birth defects²⁻⁴, giving additional emphasis to the importance of folic acid for healthy pregnancies.

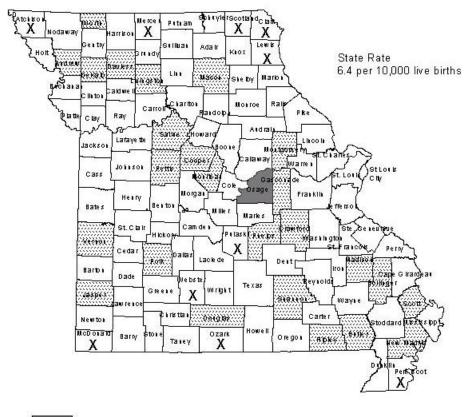
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Map 1
Births with Defects
by County of Residence
Missouri 1996-2000



Map 2 Births with Neural Tube Defects by County of Residence Missouri 1996-2000



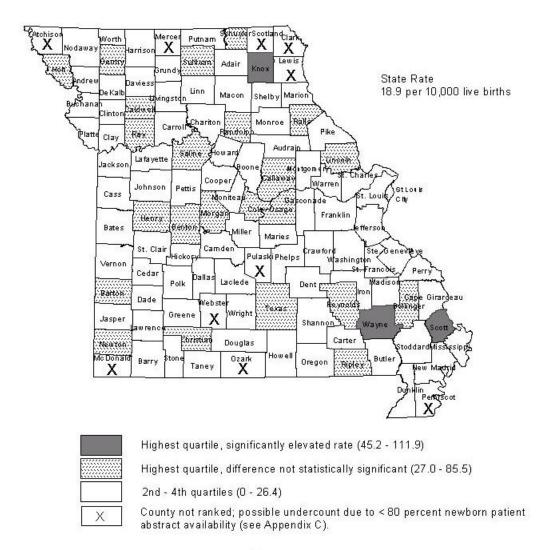
Highest quartile, significantly elevated rate (36.1)

Highest quartile, elevated rate not statistically significant (10.1 - 80.6)

2nd - 4th quartiles (0.0 - 10.0)

County not ranked; possible undercount due to < 80 percent newborn patient abstract availability (see Appendix C).

Map 3 Births with Oral Clefts by County of Residence Missouri 1996-2000



Map 4
Births with Down Syndrome
by County of Residence
Missouri 1996-2000



Highest quartile, elevated rate not statistically significant (18.6 - 46.2)

2nd - 4th quartiles (0.0 - 18.1)

County not ranked; possible undercount due to < 80 percent newborn patient abstract availability (see Appendix C).

No county had a statistically significant elevated rate for Down Syndrome

Table 1

Resident Live Births with One or More Reported Birth Defect
by Year of Birth, Maternal Race, Sex, and Maternal Age: Missouri 1996-2000

	Total Births	Births with Defects	Rate per 10,000	95% Confidence Interval
All Births	374,604	21,213	566.3	558.9 - 573.7
Year of Birth				
1996	73,728	4,046	548.8	532.4 - 565.5
1997	73,940	4,159	562.5	546.0 - 579.3
1998	75,241	4,309	572.7	556.2 - 589.5
1999	75,366	4,330	574.5	558.0 - 591.4
2000	76,329	4,369	572.4	556.0 - 589.1
Maternal Race				
White	309,279	17,227	557.0	548.9 - 565.1
Black	56,118	3,552	633.0	612.9 - 653.4
Other	8,466	384	453.6	410.2 - 500.1
Sex				
Male	191,732	13,013	678.7	667.5 - 690.1
Female	182,864	8,197	448.3	438.8 - 457.8
Maternal Age				
< 20 years	51,217	3,125	610.1	589.6 - 631.2
20-24 years	101,363	5,840	576.1	561.9 - 590.7
25-29 years	104,760	5,610	535.5	522.0 - 549.3
30-34 years	77,202	4,177	541.0	525.2 - 557.2
35-39 years	33,933	2,039	600.9	575.8 - 626.7
40 + years	6,072	418	688.4	626.0 - 755.0

Table 2 Selected Birth Defects Reported for Resident Live Births By Year of Birth: Missouri 1996-2000

		Number of Births				Rate per 10,000						
1	996-2000	1996	1997	1998	1999	2000	1996-2000	1996	1997	1998	1999	2000
TOTAL BIRTHS WITH CONGENITAL ANOMALIES	21,213	4,046	4,159	4,309	4,330	4,369	566.3	548.8-L		572.7	574.5	572.4
Central Nervous System Anomalies	1,258	282	247	270	250	209	33.6	38.2	33.4	35.9	33.2	27.4
Neural tube defects	240	60	47	47	40	46	6.4	8.1	6.4	6.2	5.3	6.0
Anencephalus	52	11	13	9	14	5	1.4	1.5	1.8	1.2	1.9	0.7
Spina bifida without anencephalus	188	49	34	38	26	41	5.0	6.6	4.6	5.1	3.4	5.4
Hydrocephalus without spina bifida	352	72	75	69	7 6	60	9.4	9.8	10.1	9.2	10.1	7.9
Microcephalus	244	54	52	56	50	32	6.5	7.3	7.0	7.4	6.6	4.2
Eye Anomalies	538	83	100	133	109	113	14.4	11.3	13.5	17.7	14.5	14.8
Anophthalmos, microphthalmos	76	13	11	18	2.3	11	2.0	1.8	1.5	2.4	3.1	1.4
Congenital cataract and lens anomalies	111	20	18	22	25	26	3.0	2.7	2.4	2.9	3.3	3.4
Ear, Face, Neck Anomalies	637	103	103	132	141	158	17.0	14.0	13.9	17.5	18.7	20.7
Heart Anomalies	5,032	1,001	986	1,080	990	975	134.3	135.8-H	133.4	143.5	131.4	127.7
Common truncus	37	10	7	9	4	7	1.0	1.4	0.9	1.2	0.5	0.9
Transposition of great vessels	166	27	40	26	42	31	4.4	3.7	5.4	3.5	5.6	4.1
Tetralogy of Fallot	216	34	47	42	36	57	5.8	4.6	6.4	5.6	4.8	7.5
Ventricular septal defect	1,665	351	324	365	331	294	44.4	47.6	43.8	48.5	43.9	38.5
Atrial septal defect	2,577	496	520	567	499	495	68.8	67.3-H	70.3	75.4	66.2	64.9
Pulmonary valve anomalies	527	116	114	100	99	98	14.1	15.7	15.4	13.3	13.1	12.8
Hypoplastic left heart syndrome	135	27	22	23	38	25	3.6	3.7	3.0	3.1	5.0	3.3
Other Circulatory and Respiratory Anomalies	4,754	871	951	994	995	943	126.9	118.1-Н	128.6	132.1	132.0	123.5
Patent ductus arteriosus	1,974	374	389	408	413	390	52.7	50.7-H	52.6	54.2	54.8	51.1
Coarctation of aorta	245	54	48	57	46	40	6.5	7.3	6.5	7.6	6.1	5.2
Pulmonary artery anomalies	711	122	153	154	136	146	19.0	16.5	20.7	20.5	18.0	19.1
Agenesis of lung	249	43	45	61	42	58	6.6	5.8	6.1	8.1	5.6	7.6
Oral Clefts	709	139	142	129	146	153	18.9	18.9	19.2	17.1	19.4	20.0
Cleft palate only	226	32	42	45	49	58	6.0	4.3-L	5.7	6.0	6.5	7.6
Cleft lip with, without cleft palate	441	104	91	72	89	85	11.8	14.1	12.3	9.6	11.8	11.1
Digestive System Anomalies	2,442	496	495	490	498	463	65.2	67.3-H	66.9	65.1	66.1	60.7
Tracheo-esophageal fistula	110	22	18	17	30	23	2.9	3.0	2.4	2.3	4.0	3.0
Small intestine atresia, stenosis	173	40	32	33	31	37	4.6	5.4	4.3	4.4	4.1	4.8
Large intestine, rectum atresia, stenosis		45	35	37	42	37	5.2	6.1	4.7	4.9	5.6	4.8
Pyloric stenosis	1,183	247	240	243	227	226	31.6	33.5-H	32.5	32.3	30.1	29.6

Table 2 continued

			Number	of Births	s		Rate per 10,000					
	1996-2000	1996	1997	1998	1999	2000	1996-2000	1996	1997	1998	1999	2000
Genital Organ Anomalies	3,763	681	752	730	765	835	100.5	92.4	101.7	97.0	101.5	109.4
Undescended testicle	1,614	289	331	334	315	345	43.1	39.2	44.8	44.4	41.8	45.2
Hypospadias and epispadias	1,634	321	340	280	334	359	43.6	43.5	46.0	37.2	44.3	47.0
Urinary Organ Anomalies	1,527	317	299	309	292	310	40.8	43.0	40.4	41.1	38.7	40.6
Renal agenesis	177	40	36	43	33	25	4.7	5.4	4.9	5.7	4.4	3.3
Obstructive urinary defects	790	154	158	157	145	176	21.1	20.9	21.4	20.9	19.2	23.1
Musculoskeletal, Integument Anomalies	5,284	997	1,011	1,048	1,094	1,134	141.1	135.2	136.7	139.3	145.2	148.6
Congenital dislocation of hip	824	177	155	144	161	187	22.0	24.0	21.0	19.1	21.4	24.5
Clubfoot	1,074	202	212	204	209	247	28.7	27.4	28.7	27.1	27.7	32.4
Reduction deformity upper limb	118	29	22	27	17	23	3.1	3.9	3.0	3.6	2.3	3.0
Reduction deformity lower limb	70	10	14	15	14	17	1.9	1.4	1.9	2.0	1.9	2.2
Skull and facial bone anomalies	1,113	188	196	200	276	253	29.7	25.5	26.5	26.6	36.6-H	33.1
Diaphragm defects	169	34	32	34	35	34	4.5	4.6	4.3	4.5	4.6	4.5
Abdominal wall defects	279	55	51	58	54	61	7.4	7.5	6.9	7.7	7.2	8.0
Integument anomalies	406	64	72	89	88	93	10.8	8.7	9.7	11.8	11.7	12.2
Chromosomal Anomalies	827	154	143	189	175	166	22.1	20.9	19.3	25.1	23.2	21.7
Trisomy 21 (Down syndrome)	512	95	82	111	116	108	13.7	12.9	11.1	14.8	15.4	14.1
Trisomy 13 (Patau syndrome)	32	4	4	13	7	4	0.9	0.5	0.5	1.7	0.9	0.5
Trisomy 18 (Edwards syndrome)	67	14	11	14	9	19	1.8	1.9	1.5	1.9	1.2	2.5
OTHER SELECTED CONDITIONS												
Congenital infections	2,397	499	460	441	540	457	64.0	67.7	62.2	58.6	71.7-H	59.9-L
Congenital pneumonia	2,072	418	388	361	486	419	55.3	56.7	52.5	48.0	64.5-H	54.9
Congenital syphilis	159	45	35	44	24	11	4.2	6.1-L	4.7	5.8	3.2	1.4
Hereditary anemias	239	40	57	54	39	49	6.4	5.4	7.7	7.2	5.2	6.4
Sickle cell anemia	112	13	25	27	21	26	3.0	1.8	3.4	3.6	2.8	3.4
Infantile cerebral palsy	176	21	44	36	48	27	4.7	2.8	6.0	4.8	6.4	3.5
Epilepsy	407	70	81	105	83	68	10.9	9.5	11.0	14.0	11.0	8.9
Congenital hypothyroidism	94	25	20	24	16	9	2.5	3.4	2.7	3.2	2.1	1.2
Intracranial hemorrhage	1,529	320	259	293	310	347	40.8	43.4-H	35.0	38.9	41.1	45.5
Total Births	374,604	73,728	73,940	75,241	75,366	76,329						

 $^{{\}tt H,L}$ Rate is significantly higher, lower than rate for preceeding year

				Rate per 10,000						
	All Race/Sex	White	Black	Male	Female	All Race/Sex	White	Black	Male	Female
TOTAL BIRTHS WITH CONGENITAL ANOMALIES	21,213	17,227	3,552	13,013	8,197	566.3	557.0-L		 678.7-Н	448.3
Central Nervous System Anomalies	1,258	995	243	656	602	33.6	32.2-L	43.3	34.2	32.9
Neural tube defects	240	214	23	118	122	6.4	6.9	4.1	6.2	6.7
Anencephalus	52	47	3	27	25	1.4	1.5	0.5	1.4	1.4
Spina bifida without anencephalus	188	167	20	91	97	5.0	5.4	3.6	4.7	5.3
Hydrocephalus without spina bifida	352	269	73	210	142	9.4	8.7-L	13.0	11.0-H	7.8
Microcephalus	244	180	62	96	148	6.5	5.8-L	11.0	5.0-L	8.1
Eye Anomalies	538	389	132	274	264	14.4	12.6-L	23.5	14.3	14.4
Anophthalmos, microphthalmos	76	60	15	39	37	2.0	1.9	2.7	2.0	2.0
Congenital cataract and lens anomalies	111	95	15	61	50	3.0	3.1	2.7	3.2	2.7
Ear, Face, Neck Anomalies	637	465	151	337	300	17.0	15.0-L	26.9	17.6	16.4
Heart Anomalies	5,032	3,921	1,007	2,691	2,340	134.3	126.8-L	179.4	140.4-H	128.0
Common truncus	37	32	5	23	14	1.0	1.0	0.9	1.2	0.8
Transposition of great vessels	166	139	25	105	61	4.4	4.5	4.5	5.5-H	3.3
Tetralogy of Fallot	216	173	35	130	86	5.8	5.6	6.2	6.8	4.7
Ventricular septal defect	1,665	1,384	254	819	846	44.4	44.7	45.3	42.7	46.3
Atrial septal defect	2,577	1,960	562	1,393	1,184	68.8	63.4-L	100.1	72.7-H	64.7
Pulmonary valve anomalies	527	385	128	282	245	14.1	12.4-L	22.8	14.7	13.4
Hypoplastic left heart syndrome	135	111	21	72	63	3.6	3.6	3.7	3.8	3.4
Other Circulatory and Respiratory Anomalies	4,754	3,775	895	2,611	2,142	126.9	122.1-L	159.5	136.2-Н	117.1
Patent ductus arteriosus	1,974	1,552	381	1,032	942	52.7	50.2-L	67.9	53.8	51.5
Coarctation of aorta	245	212	27	146	99	6.5	6.9	4.8	7.6	5.4
Pulmonary artery anomalies	711	513	182	380	331	19.0	16.6-L	32.4	19.8	18.1
Agenesis of lung	249	204	40	144	104	6.6	6.6	7.1	7.5	5.7
Oral Clefts	709	636	59	397	312	18.9	20.6-H	10.5	20.7	17.1
Cleft palate only	226	195	26	103	123	6.0	6.3	4.6	5.4	6.7
Cleft lip with, without cleft palate	441	402	31	274	167	11.8	13.0-H	5.5	14.3-H	9.1
Digestive System Anomalies	2,442	2,107	297	1,661	781	65.2	68.1-H	52.9	86.6-H	42.7
Tracheo-esophageal fistula	110	95	14	62	48	2.9	3.1	2.5	3.2	2.6
Small intestine atresia, stenosis	173	141	28	86	87	4.6	4.6	5.0	4.5	4.8
Large intestine, rectum atresia, stenosis	s 196	178	16	107	89	5.2	5.8-H	2.9	5.6	4.9
Pyloric stenosis	1,183	1,080	90	964	219	31.6	34.9-H	16.0	50.3-H	12.0

Table 3 continued

			Number -			Rate per 10,000				
	All Race/Sex	White	Black	Male	Female	All Race/Sex	White	Black	Male	Female
Genital Organ Anomalies	3,763	3,108	 583	3,532	230	100.5	100.5	103.9	 184.2-н	12.6
Undescended testicle	1,614	1,329	252	1,610		43.1	43.0	44.9	84.0	
Hypospadias and epispadias	1,634	1,385	227	1,633		43.6	44.8	40.5	85.2	
Urinary Organ Anomalies	1,527	1,260	229	1,021	505	40.8	40.7	40.8	53.3-H	27.6
Renal agenesis	177	148	25	122	54	4.7	4.8	4.5	6.4-H	3.0
Obstructive urinary defects	790	658	111	552	238	21.1	21.3	19.8	28.8-Н	13.0
Musculoskeletal, Integument Anomalies	5,284	4,415	761	2,623	2,659	141.1	142.8	135.6	136.8	145.4
Congenital dislocation of hip	824	766	46	232	592	22.0	24.8-H	8.2	12.1-L	32.4
Clubfoot	1,074	905	152	604	470	28.7	29.3	27.1	31.5-H	
Reduction deformity upper limb	118	94	20	67	51	3.1	3.0	3.6	3.5	2.8
Reduction deformity lower limb	70	54	15	43	27	1.9	1.7	2.7	2.2	1.5
Skull and facial bone anomalies	1,113	966	129	642	471	29.7	31.2-H	23.0	33.5-H	
Diaphragm defects	169	133	30	83	85	4.5	4.3	5.3	4.3	4.6
Abdominal wall defects	279	215	58	157	122	7.4	7.0	10.3	8.2	6.7
Integument anomalies	406	256	136	235	171	10.8	8.3-L	24.2	12.3	9.4
Chromosomal Anomalies	827	679	130	427	399	22.1	22.0	23.2	22.3	21.8
Trisomy 21 (Down syndrome)	512	435	66	269	243	13.7	14.1	11.8	14.0	13.3
Trisomy 13 (Patau syndrome)	32	24	7	16	16	0.9	0.8	1.2	0.8	0.9
Trisomy 18 (Edwards syndrome)	67	50	16	32	35	1.8	1.6	2.9	1.7	1.9
OTHER SELECTED CONDITIONS										
Congenital infections	2,397	1,846	499	1,485	912	64.0	59.7-L	88.9	77.5-H	49.9
Congenital pneumonia	2,072	1,709	323	1,316	756	55.3	55.3	57.6	68.6-H	41.3
Congenital syphilis	159	21	133	79	80	4.2	0.7-L	23.7	4.1	4.4
Hereditary anemias	239	87	148	140	99	6.4	2.8-L	26.4	7.3	5.4
Sickle cell anemia	112	3	109	62	50	3.0	0.1-L	19.4	3.2	2.7
Infantile cerebral palsy	176	134	41	103	73	4.7	4.3-L	7.3	5.4	4.0
Epilepsy	407	294	103	220	187	10.9	9.5-L	18.4	11.5	10.2
Congenital hypothyroidism	94	70	21	45	49	2.5	2.3	3.7	2.3	2.7
Intracranial hemorrhage	1,529	1,067	429	912	617	40.8	34.5-L	76.4	47.6-H	33.7
Total Births	374,604	309,279	56,118	191,732	182,864					

H,L Rate is significantly higher, lower for white infants in comparison with black infants, or males in comparison with females.

Table 4 Selected Birth Defects Reported for Resident Live Births By Maternal Age: Missouri 1996-2000

	Number					Rate per 10,000						
	<20	20-24	25-29	30-34	35-39	40+	<20	20-24	25-29	30-34	35-39	40+
TOTAL BIRTHS WITH CONGENITAL ANOMALIES	3,125	5,840	5,610	4,177	2,039	418	610.1-H	 576.1-Н	535.5	541.0	 600.9-Н	 688.4-Н
Central Nervous System Anomalies	208	368	300	211	146	25	40.6-H	36.3-H	28.6	27.3	43.0-H	41.2
Neural tube defects	29	70	70	42	25	4	5.7	6.9	6.7	5.4	7.4	6.6
Anencephalus	8	9	13	14	6	2	1.6	0.9	1.2	1.8	1.8	3.3
Spina bifida without anencephalus	21	61	57	28	19	2	4.1	6.0	5.4	3.6	5.6	3.3
Hydrocephalus without spina bifida	66	107	79	55	39	6	12.9-H	10.6	7.5	7.1	11.5	9.9
Microcephalus	40	71	60	41	27	5	7.8	7.0	5.7	5.3	8.0	8.2
Eye Anomalies	62	136	158	112	61	9	12.1	13.4	15.1	14.5	18.0	14.8
Anophthalmos, microphthalmos	10	11	24	18	12	1	2.0	1.1	2.3	2.3	3.5	1.6
Congenital cataract and lens anomalies	15	26	35	26	8	1	2.9	2.6	3.3	3.4	2.4	1.6
Ear, Face, Neck Anomalies	84	191	183	117	50	12	16.4	18.8	17.5	15.2	14.7	19.8
Heart Anomalies	701	1,366	1,337	993	511	122	136.9	134.8	127.6	128.6	150.6-H	200.9-H
Common truncus	2	9	14	6	3	3	0.4	0.9	1.3	0.8	0.9	4.9
Transposition of great vessels	27	40	47	35	12	5	5.3	3.9	4.5	4.5	3.5	8.2
Tetralogy of Fallot	25	52	56	50	26	7	4.9	5.1	5.3	6.5	7.7	11.5
Ventricular septal defect	210	410	460	360	168	57	41.0	40.4	43.9	46.6	49.5	93.9-H
Atrial septal defect	381	739	667	486	247	55	74.4-H	72.9-H	63.7	63.0	72.8	90.6-H
Pulmonary valve anomalies	84	146	142	104	42	9	16.4	14.4	13.6	13.5	12.4	14.8
Hypoplastic left heart syndrome	15	45	38	26	9	2	2.9	4.4	3.6	3.4	2.7	3.3
Other Circulatory and Respiratory Anomalies	616	1,284	1,283	952	512	107	120.3	126.7	122.5	123.3	150.9-Н	176.2-Н
Patent ductus arteriosus	227	529	555	407	211	45	44.3	52.2	53.0	52.7	62.2	74.1
Coarctation of aorta	28	74	65	56	15	7	5.5	7.3	6.2	7.3	4.4	11.5
Pulmonary artery anomalies	116	209	171	130	70	15	22.6-H	20.6	16.3	16.8	20.6	24.7
Agenesis of lung	29	69	66	53	32	0	5.7	6.8	6.3	6.9	9.4	0.0
Oral Clefts	95	196	200	122	80	16	18.5	19.3	19.1	15.8	23.6	26.4
Cleft palate only	32	60	65	38	26	5	6.2	5.9	6.2	4.9	7.7	8.2
Cleft lip with, without cleft palate	60	127	120	73	50	11	11.7	12.5	11.5	9.5	14.7	18.1
Digestive System Anomalies	410	721	615	446	217	33	80.1-H	71.1-Н	58.7	57.8	63.9	54.3
Tracheo-esophageal fistula	16	29	25	29	6	5	3.1	2.9	2.4	3.8	1.8	8.2
Small intestine atresia, stenosis	29	51	32	26	26	9	5.7	5.0	3.1	3.4	7.7-H	14.8-H
Large intestine, rectum atresia, stenosis		52	52	36	17	2	7.2	5.1	5.0	4.7	5.0	3.3
Pyloric stenosis	219	375	306	196	80	7	42.8-H	37.0-H	29.2	25.4	23.6	11.5-L

Table 4 continued

			Nu	mber			Rate per 10,000					
	<20	20-24	25-29	30-34	35-39	40+	<20	20-24	25-29	30-34	35-39	40+
Genital Organ Anomalies	547	1,055	1,048	739	315	59	106.8	104.1	100.0	95.7	92.8	97.2
Undescended testicle	228	471	440	310	141	24	44.5	46.5	42.0	40.2	41.6	39.5
Hypospadias and epispadias	230	453	439	333	147	32	44.9	44.7	41.9	43.1	43.3	52.7
Urinary Organ Anomalies	209	398	411	297	176	36	40.8	39.3	39.2	38.5	51.9-Н	59.3
Renal agenesis	32	57	35	30	20	3	6.2	5.6	3.3	3.9	5.9	4.9
Obstructive urinary defects	105	200	224	151	91	19	20.5	19.7	21.4	19.6	26.8	31.3
Musculoskeletal, Integument Anomalies	822	1,463	1,349	1,070	493	84	160.5-Н	144.3	128.8	138.6	145.3	138.3
Congenital dislocation of hip	97	183	241	214	70	18	18.9	18.1-L	23.0	27.7	20.6	29.6
Clubfoot	180	300	270	220	86	17	35.1-H	29.6	25.8	28.5	25.3	28.0
Reduction deformity upper limb	21	30	32	21	12	2	4.1	3.0	3.1	2.7	3.5	3.3
Reduction deformity lower limb	17	17	12	16	7	1	3.3	1.7	1.1	2.1	2.1	1.6
Skull and facial bone anomalies	160	320	296	209	121	6	31.2	31.6	28.3	27.1	35.7	9.9-L
Diaphragm defects	23	53	41	30	17	5	4.5	5.2	3.9	3.9	5.0	8.2
Abdominal wall defects	82	104	41	28	20	4	16.0-H	10.3-H	3.9	3.6	5.9	6.6
Integument anomalies	59	126	80	95	37	9	11.5	12.4	7.6	12.3	10.9	14.8
Chromosomal Anomalies	74	145	163	164	186	95	14.4	14.3	15.6	21.2	54.8-H	156.5-H
Trisomy 21 (Down syndrome)	47	79	79	109	131	67	9.2	7.8	7.5	14.1	38.6-H	110.3-H
Trisomy 13 (Patau syndrome)	5	6	8	3	9	1	1.0	0.6	0.8	0.4	2.7-H	1.6
Trisomy 18 (Edwards syndrome)	0	16	19	13	10	9	0.0-L	1.6	1.8	1.7	2.9	14.8-H
OTHER SELECTED CONDITIONS												
Congenital infections	376	768	638	394	177	44	73.4-Н	75.8-H	60.9	51.0	52.2	72.5
Congenital pneumonia	309	663	560	341	161	38	60.3-H	65.4-H	53.5	44.2	47.4	62.6
Congenital syphilis	20	53	42	32	7	5	3.9	5.2	4.0	4.1	2.1	8.2
Hereditary anemias	44	72	66	43	12	2	8.6	7.1	6.3	5.6	3.5	3.3
Sickle cell anemia	25	40	27	17	3	0	4.9	3.9	2.6	2.2	0.9	0.0
Infantile cerebral palsy	31	44	47	35	18	1	6.1	4.3	4.5	4.5	5.3	1.6
Epilepsy	68	116	99	89	32	3	13.3	11.4	9.5	11.5	9.4	4.9
Congenital hypothyroidism	14	29	14	21	14	2	2.7	2.9	1.3	2.7	4.1	3.3
Intracranial hemorrhage	290	446	343	285	135	30	56.6-H	44.0-H	32.7	36.9	39.8	49.4
Total Births	51,217	101,363	104,760	77,202	33,933	6,072						

H,L Rate is significantly higher, lower than rate for 25-34 years

Table 5 Selected Birth Defects Reported for Resident Live Births, with Number and Percent
Low Birth Weight (less than 2500 g), Preterm (less than 37 weeks), and Infant Deaths (less than one year): Missouri 1996-2000

		Nu	mber		Percent				
	Total	Low Birth Weight	Preterm	Infant Death	Low Birth Weight	Preterm	Infant Death		
TOTAL BIRTHS WITH CONGENITAL ANOMALIES	21,213	3,957	4,271	1,013	18.7	20.1	4.8		
Central Nervous System Anomalies	1,258	542	483	226	43.1	38.4	18.0		
Neural tube defects	240	81	67	66	33.8	27.9	27.5		
Anencephalus	52	43	34	52	82.7	65.4	100.0		
Spina bifida without anencephalus	188	38	33	14	20.2	17.6	7.4		
Hydrocephalus without spina bifida	352	182	188	70	51.7	53.4	19.9		
Microcephalus	244	118	77	26	48.4	31.6	10.7		
Eye Anomalies	538	162	141	41	30.1	26.2	7.6		
Anophthalmos, microphthalmos	76	22	15	10	28.9	19.7	13.2		
Congenital cataract and lens anomalies	111	16	19	4	14.4	17.1	3.6		
Ear, Face, Neck Anomalies	637	103	101	45	16.2	15.9	7.1		
Heart Anomalies	5,032	1,361	1,418	447	27.0	28.2	8.9		
Common truncus	37	9	10	12	24.3	27.0	32.4		
Transposition of great vessels	166	26	26	38	15.7	15.7	22.9		
Tetralogy of Fallot	216	48	36	27	22.2	16.7	12.5		
Ventricular septal defect	1,665	354	371	114	21.3	22.3	6.8		
Atrial septal defect	2,577	728	776	187	28.2	30.1	7.3		
Pulmonary valve anomalies	527	207	211	40	39.3	40.0	7.6		
Hypoplastic left heart syndrome	135	23	23	85	17.0	17.0	63.0		
Other Circulatory and Respiratory Anomalies	4,754	1,037	1,061	441	21.8	22.3	9.3		
Patent ductus arteriosus	1,974	*	*	113	*	*	5.7		
Coarctation of aorta	245	33	32	41	13.5	13.1	16.7		
Pulmonary artery anomalies	711	309	302	55	43.5	42.5	7.7		
Agenesis of lung	2 4 9	170	158	160	68.3	63.5	64.3		
Oral Clefts	709	124	101	48	17.5	14.2	6.8		
Cleft palate only	226	47	41	11	20.8	18.1	4.9		
Cleft lip with, without cleft palate	441	64	51	26	14.5	11.6	5.9		
Disaction Control Associate	2 442	450	F 2.4	107	10.4	21 5	4 4		
Digestive System Anomalies	2,442	450	524	107	18.4	21.5	4.4		
Tracheo-esophageal fistula	110	50	36	18	45.5	32.7	16.4		
Small intestine atresia, stenosis	173	65	78	15	37.6	45.1	8.7		
Large intestine, rectum atresia, stenosis	196	52	46	17	26.5	23.5	8.7		
Pyloric stenosis	1,183	104	168	3	8.8	14.2	0.3		

Table 5 continued

		Nu	mber			- Percent	
	Total	Low Birth Weight	Preterm	Infant Death	Low Birth Weight	Preterm	Infant Death
Genital Organ Anomalies	3,763	377	407	53	10.0	10.8	1.4
Undescended testicle	1,614	*	*	17	*	*	1.1
Hypospadias and epispadias	1,634	239	257	17	14.6	15.7	1.0
Urinary Organ Anomalies	1,527	323	353	138	21.2	23.1	9.0
Renal agenesis	177	80	67	61	45.2	37.9	34.5
Obstructive urinary defects	790	148	184	29	18.7	23.3	3.7
Musculoskeletal, Integument Anomalies	5,284	906	917	237	17.1	17.4	4.5
Congenital dislocation of hip	824	85	101	11	10.3	12.3	1.3
Clubfoot	1,074	185	198	53	17.2	18.4	4.9
Reduction deformity upper limb	118	33	19	8	28.0	16.1	6.8
Reduction deformity lower limb	70	18	16	5	25.7	22.9	7.1
Skull and facial bone anomalies	1,113	195	180	28	17.5	16.2	2.5
Diaphragm defects	169	49	46	57	29.0	27.2	33.7
Abdominal wall defects	279	114	102	40	40.9	36.6	14.3
Integument anomalies	406	46	44	6	11.3	10.8	1.5
Chromosomal Anomalies	827	262	200	146	31.7	24.2	17.7
Trisomy 21 (Down syndrome)	512	106	113	33	20.7	22.1	6.4
Trisomy 13 (Patau syndrome)	32	19	13	21	59.4	40.6	65.6
Trisomy 18 (Edwards syndrome)	67	58	24	56	86.6	35.8	83.6
OTHER SELECTED CONDITIONS							
Congenital infections	2,397	652	769	61	27.2	32.1	2.5
Congenital pneumonia	2,072	539	665	47	26.0	32.1	2.3
Congenital syphilis	159	38	39	2	23.9	24.5	1.3
Hereditary anemias	239	42	46	0	17.6	19.2	0.0
Sickle cell anemia	112	15	20	0	13.4	17.9	0.0
Infantile cerebral palsy	176	75	74	11	42.6	42.0	6.3
Epilepsy	407	75	81	26	18.4	19.9	6.4
Congenital hypothyroidism	94	47	42	3	50.0	44.7	3.2
Intracranial hemorrhage	1,529	1,222	1,225	197	79.9	80.1	12.9
Total Births	374,604	28,795	38,131	2,832	7.7	10.2	0.8

^{*} Not considered a defect in preterm or low birth weight infants

Table 6 Selected Birth Defects Reported for Resident Live Births, with Number and Percent
Participating in Medicaid (at birth), Special Health Care Needs (SHCN) and First Steps: Missouri 1996-2000

		Num	ber		Percent				
	Total	Medicaid	SHCN	First Steps	Medicaid	SHCN	First Steps		
TOTAL BIRTHS WITH CONGENITAL ANOMALIES	21,213	9,142	1,087	2,335	44.1	5.1	11.0		
Central Nervous System Anomalies	1,258	583	201	521	47.4	16.0	41.4		
Neural tube defects	240	107	48	107	45.7	20.0	44.4		
Anencephalus	52	13	0	0	25.5	0.0	0.0		
Spina bifida without anencephalus	188	94	49	108	51.4	25.9	57.6		
Hydrocephalus without spina bifida	352	168	80	191	49.6	22.9	54.3		
Microcephalus	244	129	31	96	53.5	12.6	39.5		
Eye Anomalies	538	229	51	141	43.4	9.5	26.2		
Anophthalmos, microphthalmos	76	34	5	35	45.3	6.3	46.0		
Congenital cataract and lens anomalies	111	38	11	34	35.2	9.9	30.8		
Ear, Face, Neck Anomalies	637	298	56	112	47.8	8.8	17.6		
Heart Anomalies	5,032	2,223	441	889	45.4	8.8	17.7		
Common truncus	37	14	5	11	38.9	14.8	29.6		
Transposition of great vessels	166	60	23	38	37.5	13.7	23.0		
Tetralogy of Fallot	216	90	39	64	42.9	18.1	29.7		
Ventricular septal defect	1,665	617	141	267	38.1	8.4	16.1		
Atrial septal defect	2,577	1,200	259	532	47.9	10.0	20.7		
Pulmonary valve anomalies	527	270	44	106	51.7	8.3	20.2		
Hypoplastic left heart syndrome	135	54	23	28	43.5	16.7	20.4		
Other Circulatory and Respiratory Anomalies	4,754	2,041	329	774	43.9	6.9	16.3		
Patent ductus arteriosus	1,974	822	112	248	42.5	5.7	12.6		
Coarctation of aorta	245	93	37	51	39.9	15.2	20.9		
Pulmonary artery anomalies	711	357	78	156	51.4	11.0	21.9		
Agenesis of lung	2 4 9	105	15	42	43.0	5.8	17.0		
Oral Clefts	709	280	141	198	40.9	19.8	27.9		
Cleft palate only	226	93	45	76	42.5	20.1	33.5		
Cleft lip with, without cleft palate	441	169	94	115	39.5	21.4	26.1		
Digestive System Anomalies	2,442	1,115	144	248	46.8	5.9	10.2		
Tracheo-esophageal fistula	110	45	20	35	42.9	18.2	31.8		
Small intestine atresia, stenosis	173	78	22	38	45.9	12.8	21.8		
Large intestine, rectum atresia, stenosis	196	79	25	29	42.9	12.6	14.6		
Pyloric stenosis	1,183	552	16	34	47.5	1.4	2.9		

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Table 6 continued

		Number			Percent		
	Total	Medicaid	SHCN	First Steps	Medicaid	SHCN	First Steps
Genital Organ Anomalies	3,763	1,525	 77	165	41.6	2.0	4.4
Undescended testicle	1,614	674	26	63	43.0	1.6	3.9
Hypospadias and epispadias	1,634	628	35	68	39.6	2.1	4.2
Urinary Organ Anomalies	1,527	619	77	133	41.4	5.0	8.7
Renal agenesis	177	86	14	21	49.7	8.0	11.7
Obstructive urinary defects	790	317	40	73	40.7	5.0	9.3
Musculoskeletal, Integument Anomalies	5,284	2,336	286	552	45.0	5.4	10.5
Congenital dislocation of hip	824	274	32	52	33.5	3.9	6.3
Clubfoot	1,074	486	54	118	46.2	5.0	11.0
Reduction deformity upper limb	118	60	20	45	52.2	16.9	38.2
Reduction deformity lower limb	70	40	8	12	58.8	11.7	16.7
Skull and facial bone anomalies	1,113	501	91	189	46.0	8.2	17.0
Diaphragm defects	169	77	10	21	47.2	5.9	12.6
Abdominal wall defects	279	174	20	19	63.7	7.1	6.7
Integument anomalies	406	209	9	33	52.5	2.3	8.2
Chromosomal Anomalies	827	262	112	460	33.3	13.5	55.6
Trisomy 21 (Down syndrome)	512	164	80	351	33.5	15.6	68.6
Trisomy 13 (Patau syndrome)	32	13	0	5	41.9	0.0	14.3
Trisomy 18 (Edwards syndrome)	67	22	6	13	35.5	9.4	18.9
OTHER SELECTED CONDITIONS							
Congenital infections	2,397	1,186	86	211	50.3	3.6	8.8
Congenital pneumonia	2,072	982	68	168	48.2	3.3	8.1
Congenital syphilis	159	113	1	8	72.4	0.9	5.3
Hereditary anemias	239	163	16	11	69.7	6.5	4.5
Sickle cell anemia	112	88	11	3	80.0	10.1	3.0
Infantile cerebral palsy	176	88	41	146	51.5	23.2	83.2
Epilepsy	407	199	75	248	49.8	18.4	60.8
Congenital hypothyroidism	94	51	12	33	56.7	12.8	34.6
Intracranial hemorrhage	1,529	741	176	545	49.3	11.5	35.6
Total Births	374,604	146,297	2,229	5,569	40.5	0.6	1.5

Table 7 Live Births with One or More Birth Defects by County of Residence: Missouri 1996-2000

		Rate per			Rate per			Rate per
County	Number	10,000 *	County	Number	10,000 *	County	Number	10,000 *
Missouri Total	21,213	566.3	Grundy	28	431.4	Pettis	134	479.3
Adair	98	712.2-H	Harrison	24	457.1	Phelps	166	731.6-H
Andrew	34	375.3-L	Henry	68	516.7	Pike	63	632.5
Atchison	7	243.1**	Hickory	26	718.2	Platte	261	525.4
Audrain	113	673.4	Holt	10	427.4	Polk	96	550.1
Barry	125	537.9	Howard	36	656.9	Pulaski	149	485.5**
Barton	43	471.0	Howell	99	407.2-L	Putnam	20	660.1
Bates	56	546.3	Iron	31	466.9	Ralls	36	796.5
Benton	41	532.5	Jackson	2,856	580.9	Randolph	113	686.1
Bollinger	41	602.1	Jasper	443	556.4	Ray	94	625.0
Boone	502	572.1	Jefferson	724	527.9	Reynolds	20	561.8
Buchanan	286	506.4	Johnson	148	443.1-L	Ripley	49	587.5
Butler	149	553.3	Knox	13	485.1	St Charles	1,102	543.2
Caldwell	36	651.0	Laclede	131	605.9	St Clair	23	478.2
Callaway	155	658.7	Lafayette	88	464.9	St Francois	184	539.9
Camden	146	850.8-H	Lawrence	139	572.3	St Louis City	1,638	584.1
Cape Girardeau	276	677.3-H	Lewis	33	508.5**	St Louis County	3,460	537.7-L
Carroll	22	369.7-L	Lincoln	149	584.8	Ste Genevieve	47	504.3
Carter	19	466.8	Linn	50	565.6	Saline	62	438.8
Cass	283	512.4	Livingston	39	468.8	Schuyler	21	833.3
Cedar	28	362.2-L	McDonald	43	254.6**	Scotland	14	409.4**
Chariton	25	637.8	Macon	69	726.3	Scott	181	629.8
Christian	266	718.7-H	Madison	49	714.3	Shannon	18	379.7
Clark	18	448.9**	Maries	33	652.2	Shelby	19	484.7
Clay	663	518.0-L	Marion	94	465.6	Stoddard	102	614.1
Clinton	76	627.1	Mercer	8	394.1**	Stone	111	695.1-H
Cole	331	735.9-H	Miller	107	670.8	Sullivan	32	621.4
Cooper	61	663.0	Mississippi	69	659.7	Taney	141	544.2
Crawford	95	609.4	Moniteau	45	455.9	Texas	67	508.3
Dade	35	790.1	Monroe	28	528.3	Vernon	80	559.0
Dallas	73	710.8	Montgomery	41	584.0	Warren	96	611.5
Daviess	2.3	382.7	Morgan	63	574.3	Washington	106	682.5
De Kalb	15	276.8-L	New Madrid	86	659.5	Wayne	46	626.7
Dent	75	845.5-H	Newton	202	580.5	Webster	122	523.8**
Douglas	43	622.3	Nodaway	48	459.8	Worth	5	403.2
Dunklin	147	610.7	Oregon	26	441.4	Wright	72	575.1
Franklin	406	636.0-H	Osage	50	601.7	-		
Gasconade	58	668.2	Ozark	22	427.2**			
Gentry	12	298.5-L	Pemiscot	70	382.5**			
Greene	1,016	653.8-H	Perry	76	632.8			

^{*} Rates with fewer than 20 events in numerator are unstable and should be used with caution.

^{**} Probable undercount due to low newborn patient abstract availability (see Appendix C)

H,L Significantly higher, lower rate than that for rest of state.

Table 8 Live Births with Neural Tube Defects by County of Residence: Missouri 1996-2000

		Rate per			Rate per			Rate per
County	Number	10,000 *	County	Number	10,000 *	County	Number	10,000 *
 Missouri Total	240	6.4	Grundy		0.0	Pettis	4	14.3
Adair	0	0.0	Harrison	0	0.0	Phelps	4	17.6
Andrew	1	11.0	Henry	1	7.6	Pike	1	10.0
Atchison	0	0.0**	Hickory	0	0.0	Platte	3	6.0
Audrain	1	6.0	Holt	0	0.0	Polk	2	11.5
Addiain	_	0.0	1101 C	Ü	0.0	FOIR	2	11.5
Barry	0	0.0	Howard	0	0.0	Pulaski	4	13.0**
Barton	0	0.0	Howell	2	8.2	Putnam	0	0.0
Bates	0	0.0	Iron	0	0.0	Ralls	0	0.0
Benton	0	0.0	Jackson	31	6.3	Randolph	1	6.1
Bollinger	1	14.7	Jasper	10	12.6	Ray	0	0.0
Boone	6	6.8	Jefferson	9	6.6	Reynolds	0	0.0
Buchanan	2	3.5	Johnson	1	3.0	Ripley	1	12.0
Butler	3	11.1	Knox	0	0.0	St Charles	8	3.9
Caldwell	0	0.0	Laclede	2	9.3	St Clair	0	0.0
Callaway	2	8.5	Lafayette	1	5.3	St Francois	1	2.9
Camden	1	5.8	Lawrence	0	0.0	St Louis City	21	7.5
Cape Girardeau	1	2.5	Lewis	0	0.0**	St Louis County	41	6.4
Carroll	0	0.0	Lincoln	2	7.8	Ste Genevieve	0	0.0
Carter	0	0.0	Linn	0	0.0	Saline	2	14.2
Cass	1	1.8	Livingston	2	24.0	Schuyler	0	0.0
Cedar	0	0.0	McDonald	1	5.9**	Scotland	0	0.0**
Chariton	0	0.0	McDonaid Macon	2	21.1	Scott	3	10.4
Christian	2	5.4	Madison	1	14.6	Shannon	1	21.1
Clark	0	0.0**	Madison Maries	0	0.0	Shannon Shelby	0	0.0
Clark	8	6.3	Marion	0	0.0	Stoddard	0	0.0
Clay	0	0.3	Marion	U	0.0	Stoddard	U	0.0
Clinton	0	0.0	Mercer	0	0.0**	Stone	0	0.0
Cole	4	8.9	Miller	1	6.3	Sullivan	0	0.0
Cooper	1	10.9	Mississippi	0	0.0	Taney	2	7.7
Crawford	2	12.8	Moniteau	1	10.1	Texas	1	7.6
Dade	0	0.0	Monroe	0	0.0	Vernon	3	21.0
Dallas	0	0.0	Montgomery	1	14.2	Warren	0	0.0
Daviess	1	16.6	Morgan	0	0.0	Washington	1	6.4
De Kalb	2	36.9	New Madrid	2	15.3	Wayne	0	0.0
Dent	0	0.0	Newton	3	8.6	Webster	1	4.3**
Douglas	1	14.5	Nodaway	0	0.0	Worth	1	80.6
Dunklin	0	0.0	Oregon	0	0.0	Wright	0	0.0
Franklin	4	6.3	Osage	3	36.1-Н		Ü	0.0
Gasconade	1	11.5	Ozark	0	0.0**			
Gentry	0	0.0	Pemiscot	1	5.5**			
Greene	11	7.1	Perry	1	8.3			
01 66116	11	/ . ±	LCTTÀ	1	0.5			

^{*} Rates with fewer than 20 events in numerator are unstable and should be used with caution.

^{**} Possible undercount due to low newborn patient abstract availability (see Appendix C)

H Significantly higher rate than that for rest of state.

Table 9 Live Births with Oral Clefts by County of Residence: Missouri 1996-2000

		Rate per			Rate per			Rate per
County	Number	10,000 *	County	Number	10,000 *	County	Number	10,000 *
Missouri Total	709	18.9	Grundy	0	0.0	Pettis	3	10.7
Adair	3	21.8	Harrison	0	0.0	Phelps	2	8.8
Andrew	2	22.1	Henry	4	30.4	Pike	1	10.0
Atchison	0	0.0**	Hickory	0	0.0	Platte	12	24.2
Audrain	4	23.8	Holt	2	85.5	Polk	4	22.9
Barry	3	12.9	Howard	1	18.2	Pulaski	4	13.0**
Barton	4	43.8	Howell	6	24.7	Putnam	0	0.0
Bates	1	9.8	Iron	1	15.1	Ralls	2	44.2
Benton	4	51.9	Jackson	97	19.7	Randolph	6	36.4
Bollinger	2	29.4	Jasper	12	15.1	Ray	6	39.9
Boone	21	23.9	Jefferson	23	16.8	Reynolds	1	28.1
Buchanan	13	23.0	Johnson	5	15.0	Ripley	4	48.0
Butler	3	11.1	Knox	3	111.9-Н	St Charles	43	21.2
Caldwell	3	54.2	Laclede	4	18.5	St Clair	0	0.0
Callaway	9	38.2	Lafayette	2	10.6	St Francois	6	17.6
Camden	2	11.7	Lawrence	4	16.5	St Louis City	32	11.4
Cape Girardeau	9	22.1	Lewis	3	46.2**	St Louis County	92	14.3
Carroll	1	16.8	Lincoln	10	39.2	Ste Genevieve	1	10.7
Carter	0	0.0	Linn	1	11.3	Saline	7	49.5
Cass	13	23.5	Livingston	1	12.0	Schuyler	1	39.7
Cedar	0	0.0	McDonald	2	11.8**	Scotland	2	58.5**
Chariton	0	0.0	Macon	1	10.5	Scott	13	45.2-H
Christian	10	27.0	Madison	0	0.0	Shannon	1	21.1
Clark	0	0.0 * *	Maries	0	0.0	Shelby	1	25.5
Clay	22	17.2	Marion	4	19.8	Stoddard	3	18.1
Clinton	0	0.0	Mercer	1	49.3**	Stone	3	18.8
Cole	14	31.1	Miller	2	12.5	Sullivan	2	38.8
Cooper	2	21.7	Mississippi	2	19.1	Taney	0	0.0
Crawford	3	19.2	Moniteau	2	20.3	Texas	4	30.3
Dade	1	22.6	Monroe	0	0.0	Vernon	3	21.0
Dallas	2	19.5	Montgomery	0	0.0	Warren	2	12.7
Daviess	1	16.6	Morgan	5	45.6	Washington	4	25.8
De Kalb	1	18.5	New Madrid	3	23.0	Wayne	5	68.1-H
Dent	2	22.5	Newton	12	34.5	Webster	3	12.9**
Douglas	0	0.0	Nodaway	0	0.0	Worth	0	0.0
Dunklin	2	8.3	Oregon	1	17.0	Wright	1	8.0
Franklin	10	15.7	Osage	3	36.1			
Gasconade	2	23.0	Ozark	1	19.4**			
Gentry	2	49.8	Pemiscot	3	16.4**			
Greene	41	26.4	Perry	2	16.7			

^{*} Rates with fewer than 20 events in numerator are unstable and should be used with caution.

^{**} Possible undercount due to low newborn patient abstract availability (see Appendix C)

H Significantly higher rate than that for rest of state.

Table 10 Live Births with Down Syndrome by County of Residence: Missouri 1996-2000

County	Number	Rate per 10,000 *	County	Number	Rate per 10,000 *	County	Number	Rate per 10,000 *
Missouri Total	512	13.7	 Grundv	3	46.2	Pettis	3	10.7
Adair	4	29.1	Harrison	0	0.0	Phelps	2	8.8
Andrew	2	22.1	Henry	0	0.0	Pike	1	10.0
Atchison	2	69.4**	Hickory	0	0.0	Platte	8	16.1
Audrain	3	17.9	Holt	0	0.0	Polk	1	5.7
Barry	6	25.8	Howard	1	18.2	Pulaski	3	9.8**
Barton	0	0.0	Howell	2	8.2	Putnam	0	0.0
Bates	2	19.5	Iron	1	15.1	Ralls	1	22.1
Benton	2	26.0	Jackson	58	11.8	Randolph	2	12.1
Bollinger	0	0.0	Jasper	11	13.8	Ray	1	6.6
Boone	9	10.3	Jefferson	25	18.2	Reynolds	0	0.0
Buchanan	11	19.5	Johnson	5	15.0	Ripley	1	12.0
Butler	3	11.1	Knox	1	37.3	St Charles	36	17.7
Caldwell	0	0.0	Laclede	1	4.6	St Clair	0	0.0
Callaway	2	8.5	Lafayette	0	0.0	St Francois	4	11.7
Camden	3	17.5	Lawrence	2	8.2	St Louis City	36	12.8
Cape Girardeau	5	12.3	Lewis	1	15.4**	St Louis County	84	13.1
Carroll	1	16.8	Lincoln	6	23.5	Ste Genevieve	1	10.7
Carter	0	0.0	Linn	1	11.3	Saline	0	0.0
Cass	5	9.1	Livingston	1	12.0	Schuyler	0	0.0
Cedar	0	0.0	McDonald	0	0.0**	Scotland	0	0.0**
Chariton	1	25.5	Macon	1	10.5	Scott	5	17.4
Christian	9	24.3	Madison	0	0.0	Shannon	0	0.0
Clark	0	0.0**	Maries	1	19.8	Shelby	0	0.0
Clay	28	21.9	Marion	3	14.9	Stoddard	3	18.1
Clinton	1	8.3	Mercer	0	0.0**	Stone	6	37.6
Cole	3	6.7	Miller	2	12.5	Sullivan	0	0.0
Cooper	0	0.0	Mississippi	1	9.6	Taney	4	15.4
Crawford	1	6.4	Moniteau	3	30.4	Texas	0	0.0
Dade	0	0.0	Monroe	0	0.0	Vernon	3	21.0
Dallas	1	9.7	Montgomery	1	14.2	Warren	4	25.5
Daviess	0	0.0	Morgan	0	0.0	Washington	4	25.8
De Kalb	0	0.0	New Madrid	0	0.0	Wayne	1	13.6
Dent	4	45.1	Newton	5	14.4	Webster	1	4.3**
Douglas	0	0.0	Nodaway	1	9.6	Worth	0	0.0
Dunklin	1	4.2	Oregon	2	34.0	Wright	3	24.0
Franklin	10	15.7	Osage	1	12.0			
Gasconade	2	23.0	Ozark	0	0.0**			
Gentry	0	0.0	Pemiscot	3	16.4**			
Greene	29	18.7	Perry	1	8.3			

^{*} Rates with fewer than 20 events in numerator are unstable and should be used with caution.

^{**} Possible undercount due to low newborn patient abstract availability (see Appendix C)

H Significantly higher rate than that for rest of state.

Table 11 Selected Birth Defects Reported for Resident Live Births by County of Residence: Missouri 1996-2000

	Adair	Andrew	Atchison*	Audrain	Barry	Barton	Bates	Benton
TOTAL WITH CONGENITAL ANOMALIES	98	34	7	113	125	43	56	41
Central Nervous System	3	1	1	9	11	3	4	0
Eye	3	1	0	3	4	1	3	3
Ear, Face, Neck	1	1	0	1	6	1	4	2
Heart Anomalies	17	15	2	21	34	9	13	9
Other Circulatory, Respiratory	20	3	1	25	22	9	6	8
Oral Clefts	3	2	0	4	3	4	1	4
Digestive System	12	8	0	14	9	2	13	5
Genital Organ	16	3	1	18	22	8	13	8
Urinary Organ	5	1	0	4	5	4	2	3
Musculoskeletal, Integument	37	3	2	38	40	11	11	10
Chromosomal Anomalies	4	3	2	4	8	0	3	2
Total Births	1,376	906	288	1,678	2,324	913	1,025	770

	Bollinger	Boone	Buchanan	Butler	Caldwell	Callaway	Camden	Cape Girardeau
TOTAL WITH CONGENITAL ANOMALIES	41	502	286	149	36	155	146	276
Central Nervous System	6	29	12	10	2	5	6	12
Eye	0	10	5	5	1	2	1	8
Ear, Face, Neck	1	14	6	7	0	7	1	4
Heart Anomalies	9	94	81	25	12	38	16	42
Other Circulatory, Respiratory	9	157	47	29	9	41	30	42
Oral Clefts	2	21	13	3	3	9	2	9
Digestive System	5	47	46	26	2	23	10	41
Genital Organ	9	68	45	22	8	18	29	57
Urinary Organ	6	27	19	12	1	10	10	29
Musculoskeletal, Integument	14	123	84	45	8	38	56	76
Chromosomal Anomalies	1	15	17	5	1	4	5	8
Total Births	681	8,774	5,648	2,693	553	2,353	1,716	4,075

^{*} Possible undercount due to <80 percent patient abstract completeness (see Appendix C)

Table 11 Continued

	Carroll	Carter	Cass	Cedar	Chariton	Christian	Clark*	Clay
TOTAL WITH CONGENITAL ANOMALIES	22	19	283	28	25	266	18	663
Central Nervous System	0	2	10	1	0	15	0	30
Eye	0	0	4	0	0	5	2	15
Ear, Face, Neck	0	0	5	0	2	10	0	13
Heart Anomalies	5	5	64	6	6	78	4	156
Other Circulatory, Respiratory	6	4	52	5	8	62	3	136
Oral Clefts	1	0	13	0	0	10	0	22
Digestive System	3	1	40	3	3	15	1	73
Genital Organ	5	0	72	5	2	51	5	155
Urinary Organ	0	1	12	1	2	18	1	37
Musculoskeletal, Integument	6	8	60	10	6	66	5	147
Chromosomal Anomalies	1	0	7	0	1	12	0	35
Total Births	595	407	5,523	773	392	3,701	401	12,800
	Clinton	Cole	Cooper	Crawford	Dade	Dallas	Daviess	De Kalb
TOTAL WITH CONGENITAL ANOMALIES	76	331	61	95	35	73	23	15
Central Nervous System	0	15	5	9	1	5	1	3
Eye	0	6	0	1	0	0	1	1
Ear, Face, Neck	1	16	4	4	1	1	2	0
Heart Anomalies	18	73	13	31	11	18	3	3
Other Circulatory, Respiratory	16	61	15	23	10	19	4	2
Oral Clefts	0	14	2	3	1	2	1	1
Digestive System	10	38	3	10	1	9	8	1
Genital Organ	13	57	11	18	7	9	3	2
Urinary Organ	5	23	7	2	1	4	2	1
Musculoskeletal, Integument	22	86	19	24	8	26	5	3
Chromosomal Anomalies	2	4	1	1	0	3	0	0
Total Births	1,212	4,498	920	1,559	443	1,027	601	542

^{*} Possible undercount due to <80 percent patient abstract completeness (see Appendix C)

Table 11 Continued

	Dent	Douglas	Dunklin	Franklin	Gasconade	Gentry	Greene	Grundy
TOTAL WITH CONGENITAL ANOMALIES	75	43	147	406	58	12	1,016	28
Central Nervous System	3	3	8	23	2	0	48	2
Eye	1	0	7	11	0	0	17	1
Ear, Face, Neck	4	2	6	12	0	0	28	0
Heart Anomalies	17	16	44	102	13	4	329	8
Other Circulatory, Respiratory	15	7	23	96	8	2	261	3
Oral Clefts	2	0	2	10	2	2	41	0
Digestive System	5	2	19	56	10	3	87	6
Genital Organ	11	4	25	65	6	1	158	3
Urinary Organ	3	1	13	21	4	1	55	1
Musculoskeletal, Integument	26	14	35	102	20	5	218	9
Chromosomal Anomalies	4	1	3	11	2	0	54	3
Total Births	887	691	2,407	6,384	868	402	15,540	649
	Harrison	Henry	Hickory	Holt	Howard	Howell	Iron	Jackson
TOTAL WITH CONGENITAL ANOMALIES	24	68	26	10	36	99	31	2,856
Central Nervous System	2	4	1	0	2	9	3	147
Eye	0	0	0	1	0	5	0	70
Ear, Face, Neck	0	1	1	0	0	2	0	89
Heart Anomalies	7	18	10	2	6	24	7	718
Other Circulatory, Respiratory	4	15	7	3	9	20	9	605
Oral Clefts	0	4	0	2	1	6	1	97
Digestive System	0	11	2	2	7	14	3	290
Genital Organ	7	12	2	3	6	19	2	544
Urinary Organ	1	5	2	0	2	6	4	164
Musculoskeletal, Integument	6	17	11	3	10	20	10	790
Chromosomal Anomalies	0	0	0	0	1	6	2	101
Total Births	525	1,316	362	234	548	2,431	664	49,169

 $^{^{\}star}$ Possible undercount due to <80 percent patient abstract completeness (see Appendix C)

Table 11 Continued

	Jasper	Jefferson	Johnson	Knox	Laclede	Lafayette	Lawrence	Lewis*
TOTAL WITH CONGENITAL ANOMALIES	443	724	148	13	131	88	139	33
Central Nervous System	38	48	9	1	9	5	5	1
Eye	7	20	2	0	3	3	1	0
Ear, Face, Neck	11	23	5	1	1	1	2	2
Heart Anomalies	99	126	39	5	29	18	45	5
Other Circulatory, Respiratory	82	145	24	2	32	16	39	12
Oral Clefts	12	23	5	3	4	2	4	3
Digestive System	62	100	19	3	16	11	16	4
Genital Organ	99	142	25	0	27	16	21	6
Urinary Organ	39	60	9	0	9	8	9	1
Musculoskeletal, Integument	94	188	36	1	36	26	35	4
Chromosomal Anomalies	18	34	8	1	4	0	4	1
Total Births	7,962	13,716	3,340	268	2,162	1,893	2,429	649
	Lincoln	Linn	Livingston	McDonald*	Macon	Madison	Maries	Marion
TOTAL WITH CONGENITAL ANOMALIES	149	50	39	43	69	49	33	94
Central Nervous System	10	4	5	3	6	6	1	5
Eye	8	1	1	0	3	2	0	1
Ear, Face, Neck	6	0	1	0	2	3	1	5
Heart Anomalies	36	11	9	18	11	6	9	31
Other Circulatory, Respiratory	35	11	8	18	15	7	3	25
Oral Clefts	10	1	1	2	1	0	0	4
Digestive System	17	5	5	5	6	6	5	5
Genital Organ	28	1	8	5	9	11	7	16
Urinary Organ	13	3	1	3	6	3	0	7
Musculoskeletal, Integument	34	16	13	7	18	12	7	19
Chromosomal Anomalies	7	1	2	0	3	1	1	5
Total Births	2,548	884	832	1,689	950	686	506	2,019

^{*} Possible undercount due to <80 percent patient abstract completeness (see Appendix C)

Table 11 Continued

	Mercer*	Miller	Mississippi	Moniteau	Monroe	Montgomery	Morgan	New Madrid
TOTAL WITH CONGENITAL ANOMALIES	8	107	69	45	28	41	63	86
Central Nervous System	0	4	3	1	1	1	4	7
Eye	0	1	3	0	0	2	1	2
Ear, Face, Neck	1	3	3	0	0	2	0	1
Heart Anomalies	0	17	19	6	9	5	13	9
Other Circulatory, Respiratory	0	18	16	11	10	9	19	16
Oral Clefts	1	2	2	2	0	0	5	3
Digestive System	0	13	8	5	1	3	8	14
Genital Organ	2	21	13	8	4	7	14	16
Urinary Organ	0	5	5	3	2	1	6	5
Musculoskeletal, Integument	4	38	16	17	4	13	15	27
Chromosomal Anomalies	0	4	6	4	1	1	0	0
Total Births	203	1,595	1,046	987	530	702	1,097	1,304
	Newton	Nodaway	Oregon	Osage	Ozark*	Pemiscot*	Perry	Pettis
TOTAL WITH CONGENITAL ANOMALIES	202	48	26	50	22	70	76	134
Central Nervous System	14	3	3	5	1	7	5	11
Eye	4	0	1	1	0	2	2	5
Ear, Face, Neck	6	0	0	2	0	0	1	1
Heart Anomalies	48	10	6	10	6	26	11	39
Other Circulatory, Respiratory	37	7	6	7	6	10	18	35
Oral Clefts	12	0	1	3	1	3	2	3
Digestive System	31	3	3	7	6	4	8	16
Genital Organ	39	12	4	8	3	10	22	22
Urinary Organ	19	4	1	3	0	3	4	10
Musculoskeletal, Integument	34	15	4	12	3	10	23	23
Chromosomal Anomalies	6	2	2	2	0	4	1	3
Total Births	3,480	1,044	589	831	515	1,830	1,201	2,796

^{*} Possible undercount due to <80 percent patient abstract completeness (see Appendix C)

Table 11 Continued

	Phelps	Pike	Platte	Polk	Pulaski*	Putnam	Ralls	Randolph
TOTAL WITH CONGENITAL ANOMALIES	166	63	261	96	149	20	36	113
Central Nervous System	10	5	13	4	12	1	3	10
Eye	2	3	3	1	2	0	1	5
Ear, Face, Neck	0	1	13	1	1	0	2	3
Heart Anomalies	39	11	59	19	39	4	9	15
Other Circulatory, Respiratory	54	17	66	25	40	6	16	22
Oral Clefts	2	1	12	4	4	0	2	6
Digestive System	19	12	29	8	25	2	7	12
Genital Organ	23	9	62	12	15	1	4	14
Urinary Organ	7	6	15	7	7	1	0	8
Musculoskeletal, Integument	45	24	67	30	31	6	3	29
Chromosomal Anomalies	2	2	14	2	5	0	1	2
Total Births	2,269	996	4,968	1,745	3,069	303	452	1,647
	Ray	Reynolds	Ripley	St Charles	St Clair	St Francois	St Louis City	St Louis County
TOTAL WITH CONGENITAL ANOMALIES	94	20	49	1,102	23	184	1,638	3,460
Central Nervous System	5	1	2	51	0	10	121	234
Eye	1	0	1	36	0	4	63	100
Ear, Face, Neck	1	0	0	22	1	2	85	122
Heart Anomalies	26	6	10	206	6	30	411	803
Other Circulatory, Respiratory	19	5	11	239	6	28	400	814
Oral Clefts	6	1	4	43	0	6	32	92
Digestive System	11	3	5	145	4	41	158	375
Genital Organ	16	3	7	202	3	23	279	593
Urinary Organ	3	3	4	113	0	11	147	310
Musculoskeletal, Integument	27	5	16	267	6	61	350	787
Chromosomal Anomalies	3	0	3	58	0	6	62	154
Total Births	1,504	356	834	20,289	481	3,408	28,043	64,354

 $^{^{\}star}$ Possible undercount due to <80 percent patient abstract completeness (see Appendix C)

Table 11 Continued

	Ste Genevieve	Saline	Schuyler	Scotland*	Scott	Shannon	Shelby	Stoddard
TOTAL WITH CONGENITAL ANOMALIES	47	62	21	14	181	18	19	102
Central Nervous System	1	9	1	0	16	1	0	4
Eye	1	2	1	0	6	3	1	2
Ear, Face, Neck	1	3	0	0	6	1	1	2
Heart Anomalies	10	8	4	4	38	8	3	30
Other Circulatory, Respiratory	12	9	7	5	33	4	2	21
Oral Clefts	1	7	1	2	13	1	1	3
Digestive System	7	10	0	2	25	3	4	24
Genital Organ	8	13	2	1	35	2	4	16
Urinary Organ	4	6	4	0	18	0	2	9
Musculoskeletal, Integument	14	13	9	7	47	4	3	22
Chromosomal Anomalies	1	1	0	0	9	0	0	3
Total Births	932	1,413	252	342	2,874	474	392	1,661
	Stone	Sullivan	Taney	Texas	Vernon	Warren	Washington	Wayne
TOTAL WITH CONGENITAL ANOMALIES	Stone	Sullivan 32	Taney	Texas	Vernon 80	Warren 96	Washington	Wayne 46
TOTAL WITH CONGENITAL ANOMALIES Central Nervous System			-	67 4			-	-
Central Nervous System	111 7 4	32 2 0	141 4 5	67 4 3	80 8 1	96 4 9	106 7 1	46 3 1
Central Nervous System	111	32 2	141	67 4	80	96 4	106 7	46
Central Nervous System Eye Ear, Face, Neck Heart Anomalies	111 7 4 7	32 2 0 0	141 4 5 4 39	67 4 3 2	80 8 1 4	96 4 9 1 21	106 7 1 1 23	46 3 1 1
Central Nervous System Eye Ear, Face, Neck Heart Anomalies Other Circulatory, Respiratory	111 7 4 7 41 39	32 2 0 0 7 5	141 4 5 4 39 32	67 4 3 2 21 18	80 8 1 4 21 16	96 4 9 1 21 20	106 7 1 1 23 25	46 3 1 1 11
Central Nervous System Eye Ear, Face, Neck Heart Anomalies Other Circulatory, Respiratory Oral Clefts	111 7 4 7 41 39 3	32 2 0 0 7 5 2	141 4 5 4 39 32 0	67 4 3 2 21 18 4	80 8 1 4 21 16 3	96 4 9 1 21 20 2	106 7 1 1 23 25 4	46 3 1 1 1 11 10 5
Central Nervous System Eye Ear, Face, Neck Heart Anomalies Other Circulatory, Respiratory	111 7 4 7 41 39	32 2 0 0 7 5	141 4 5 4 39 32	67 4 3 2 21 18	80 8 1 4 21 16	96 4 9 1 21 20	106 7 1 1 23 25	46 3 1 1 11
Central Nervous System Eye Ear, Face, Neck Heart Anomalies Other Circulatory, Respiratory Oral Clefts Digestive System Genital Organ	111 7 4 7 41 39 3 12	32 2 0 0 7 5 2 2	141 4 5 4 39 32 0 10	67 4 3 2 21 18 4 11	80 8 1 4 21 16 3 12	96 4 9 1 21 20 2 7	106 7 1 1 23 25 4 18	46 3 1 1 1 11 10 5 5
Central Nervous System Eye Ear, Face, Neck Heart Anomalies Other Circulatory, Respiratory Oral Clefts Digestive System Genital Organ Urinary Organ	111 7 4 7 41 39 3 12	32 2 0 0 7 5 2 2 2	141 4 5 4 39 32 0 10 29 11	67 4 3 2 21 18 4 11 8 4	80 8 1 4 21 16 3 12 16 7	96 4 9 1 21 20 2 7	106 7 1 1 23 25 4 18 18	11 11 10 5 5
Central Nervous System Eye Ear, Face, Neck Heart Anomalies Other Circulatory, Respiratory Oral Clefts Digestive System Genital Organ Urinary Organ Musculoskeletal, Integument	111 7 4 7 41 39 3 12 22 5 25	32 2 0 0 7 5 2 2 2 5 1	141 4 5 4 39 32 0 10 29 11 36	67 4 3 2 21 18 4 11 8 4 25	80 8 1 4 21 16 3 12 16 7 11	96 4 9 1 21 20 2 7 20 10 17	106 7 1 1 23 25 4 18 18 11 26	46 3 1 1 11 10 5 5 5
Central Nervous System Eye Ear, Face, Neck Heart Anomalies Other Circulatory, Respiratory Oral Clefts Digestive System Genital Organ Urinary Organ	111 7 4 7 41 39 3 12	32 2 0 0 7 5 2 2 2	141 4 5 4 39 32 0 10 29 11	67 4 3 2 21 18 4 11 8 4	80 8 1 4 21 16 3 12 16 7	96 4 9 1 21 20 2 7	106 7 1 1 23 25 4 18 18	11 11 10 5 5

 $^{^{\}star}$ Possible undercount due to <80 percent patient abstract completeness (see Appendix C)

Table 11 Continued

	Webster*	Worth	Wright
TOTAL WITH CONGENITAL ANOMALIES	122	5	72
Central Nervous System	5	1	4
Eye	2	0	0
Ear, Face, Neck	2	0	3
Heart Anomalies	37	2	24
Other Circulatory, Respiratory	34	0	24
Oral Clefts	3	0	1
Digestive System	13	1	6
Genital Organ	30	0	10
Urinary Organ	5	0	4
Musculoskeletal, Integument	22	1	20
Chromosomal Anomalies	1	0	5
Total Births	2,329	124	1,252

^{*} Possible undercount due to <80 percent patient abstract completeness (see Appendix C)

Appendix A

The Missouri Birth Defects Registry

I. Sources of Data and Methods of Ascertaining Birth Defects.

The Missouri Department of Health and Senior Services (DHSS) birth defects registry for live born Missouri residents includes defects diagnosed within the first year of life. The registry is passive and multi-source. Data are population-based and collected statewide, although some data sources vary in completeness by area of residence. For data collection, birth defects are defined as all diagnoses listed in the congenital anomalies section of the International Classification of Diseases-9th Revision (ICD-9) and other selected conditions, primarily of genetic or prenatal origin. For most analyses, including those discussed in this proposal, the term 'birth defect' refers to diagnoses included in ICD-9 codes 740-759, with certain exclusions listed in Appendix C.

Birth defects data are available for births since 1980. Within this time span, however, there have been changes in data sources available for inclusion and in methodology of data set development. The following description of the registry applies to births since 1993. Table A-1 demonstrates the incremental contribution of birth defects cases from each data component described below to the total number of birth defect cases. In Table A-2, the number of defects, percent reported in each component, and percent reported in multiple components are listed for selected defects.

Birth Certificates. The birth defects registry is birth certificate based. It is the record to which all other data are linked. All data items recorded on the certificate are included in the registry. Birth certificates for births to Missouri residents occurring out of state are included. The birth certificate is itself a source of birth defect data. A check-box format is used, with space for hand entering (written) defects not included in the check-box list; this format follows the U.S. standard certificate.

Overall, birth certificates contribute only 16.2 percent of the total number of birth defects reported on the registry (Table A-1). The percentage of defects reported on birth certificates is much higher for more severe and readily identified defects. Birth certificate data included over 50 percent of the total number of cases of spina bifida, oral clefts, diaphragm defects, and Down syndrome reported to the registry from 1996 through 2000 (Table A-2). Because many of the birth certificate defect check-boxes cover broad categories, defect data obtained from birth certificates are often less specific than data from other sources. For example, all heart anomalies are included in a single check box, thus birth certificate data can be included in the major subcategory of heart defects, but not in more specific subcategories such as common truncus or ventricular septal defect. Appendix B includes the birth certificate availability of defect categories presented in the report.

Table A-1

Increments in Reporting of Infants with Birth Defects Obtained by the Addition of Each Data Component: Missouri 1996-2000

	Number	Incremental Contribution to Registry (%)	Incremental Rate per 10,000 Births
Birth Certificate	3,538	16.2%	91.9
Birth Plus Death Certificate	3,794	17.3%	98.0
Birth, Death and Newborn Patient Abstract	15,366	71.7%	405.8
Above Plus Program Data	15,703	73.2%	414.5
Above Plus Pediatric Patient Abstract	21,213	100.0%	566.3

Death Certificates. Another source of birth defect data is cause of death as listed on infant death certificates. Death certificates are available for nearly all infant deaths, including those occurring out of state. Virtually 100 percent of infant death records are linked to the appropriate birth certificate. Linked birth and death data are maintained in a computerized database and were added to the registry by year of birth and birth certificate number. Deaths from all causes occurring before the first birthday were incorporated into the registry. Data items included are cause of death and age at death.

Since 1999, death certificates have been coded using the 10th Revision of the International Classification of Diseases. ICD-10 codes were reconciled with the ICD-9-coded diagnoses used for the other data components to produce this report.

Birth defects were listed as the cause of 23 percent of Missouri infant deaths from 1996-2000. An additional 13 percent of infant deaths occurred among infants with birth defects but other listed causes (e.g., prematurity or respiratory distress syndrome). The data are limited in the type of defect represented because many severe birth defects have much higher morbidity than mortality. Underreporting of birth defects as a cause of death is likely inasmuch as less than 50 percent of Missouri resident infant deaths are autopsied. In addition, the causes of death recorded on the death certificate may differ from the causes determined from autopsy. In terms of ascertainment, death certificates contribute only an additional 1.1 percent of total birth defects reported over those reported on the birth certificate.

Newborn Patient Abstracts. A major source of data on both major and minor birth defects is the newborn patient abstract. The newborn patient abstract represents the initial hospitalization and is obtained from the delivery hospital. All other admissions, including immediate transfers to referral centers, are categorized as pediatric admissions and are discussed separately below. Prior to 1993 reporting was to varying degrees incomplete and patient names were not available for linking with the birth data set. A 1992 Missouri law made submission of patient abstracts mandatory for all non-federal Missouri hospitals. Hospitals are required to submit abstracts to DHSS quarterly within five months of the end of the quarter. DHSS also receives patient abstract data on Missouri residents discharged from some Illinois, Iowa, and Kansas hospitals, improving coverage for some border counties with a majority of births occurring in out-of-state hospitals. Nonetheless, there is significant under-representation of some counties' births in the patient abstract data set. Appendix C lists the percentage of each county's births with linked birth certificates and newborn abstracts.

Newborn patient abstracts are linked to the birth certificate by computer algorithm and follow-up visual examination of unlinked and multiple potential links. Using patient name, date and hospital of birth, sex, race, county and zip code, 94 percent of resident live births for 1996-2000 were linked to the appropriate newborn patient abstract. Births not linked to a newborn patient abstract primarily represent Missouri resident births occurring in military or out-of-state hospitals. Data items included in the registry are ICD-9-CM-coded diagnoses, length of stay, and total hospital charges. In terms of sequential registry development, newborn abstracts increase the percentage of birth defects from 17.3 to 71.7 percent of all birth defects reported to the registry (Table A-1); however, the relative gain is smaller for more severe birth defects since a higher percentage of these are reported on birth certificates (Table A-2).

DHSS and Department of Mental Health Program Data Bases. Also included in the registry is the administrative data base maintained for a number of DHSS programs including the following: 1) Children with Special Health Care Needs (CSHCN); 2) First Steps - Missouri's early intervention program for developmental disabilities program (Part C of the Individuals with Disabilities Act); 3) Healthy Children and Youth (EPSDT screenings); and 4) specific programs which provide services for individuals with cystic fibrosis, sickle cell anemia, hemophilia, and inborn metabolic errors requiring special diets. Of these the CSHCN and First Steps are the most important source of data on birth defects. CSHCN supports treatment for children with various birth defects from low-income families. The First Steps program provides services to children at risk for developmental delay regardless of family income. The Department of Mental Health (DMH) also serves First Steps clients and provided data on its First Steps clients for inclusion in the registry.

Both the DHSS and DMH maintain the most current information on children enrolled in the programs specified above. Discontinued clients are not dropped from the database. Linkage is attempted for all infants enrolled in any of the above-mentioned programs prior to the first birthday. A 91 percent linkage rate is achieved through computer

Table A-2 Source of Birth Defects Reported for Resident Live Births: Missouri 1996-2000

	Number	Percent by Source							
	All Sources	Multiple Sources	Birth Cert	Death Cert		Pediatric Abstract	Program Data*		
BIRTHS WITH ONE OR MORE DEFECTS	21,213	26.7	16.2	2.8	66.5	47.7	6.3		
Central Nervous System Anomalies	1,258	33.9	27.6	7.1	45.4	60.9	20.7		
Spina bifida without anencephalus	188	66.7	61.0	0.0	61.9	74.3	47.6		
Eye Anomalies	538	15.5	11.3	0.0	38.0	65.1	4.8		
Heart Anomalies	5,032	22.2	13.2	3.5	56.0	54.4	5.2		
Tetralogy of Fallot	216	40.0	NA	1.5	45.9	83.0	15.6		
Ventricular septal defect	1,665	12.6	NA	0.4	59.7	49.6	3.9		
Atrial septal defect	2,577	4.2	NA	0.1	39.3	63.2	1.7		
Other Circulatory and Respiratory Anomalies	4,754	10.9	5.8	3.3	46.7	54.6	1.7		
Patent ductus arteriosus	1,974	4.7	NA	0.0	58.1	45.6	1.1		
Oral Clefts	709	74.3	60.0	0.2	78.5	74.3	21.3		
Digestive System Anomalies	2,442	13.3	13.5	0.3	24.1	79.0	2.5		
Tracheo-esophageal fistula	110	42.9	45.7	0.0	48.6	61.4	5.7		
Pyloric stenosis	1,183	0.6	NA	0.0	1.9	98.1	0.6		
Genital Organ Anomalies	3,763	27.2	8.7	0.0	81.6	38.8	0.2		
Urinary Organ Anomalies	1,527	27.0	28.0	2.2	60.7	42.4	1.0		
Musculoskeletal, Integument Anomalies	5,284	19.6	7.2	0.9	77.4	35.4	4.2		
Congenital dislocation of hip	824	5.1	NA	0.0	87.6	16.5	1.2		
Clubfoot	1,074	43.3	28.0	0.0	77.7	48.2	2.6		
Diaphragm defects	169	53.4	51.5	2.9	56.3	69.9	3.9		
Abdominal wall defects	279	39.9	NA	1.7	89.0	46.8	6.9		
Chromosomal Anomalies	827	69.1	47.0	9.6	67.4	55.5	44.0		
Trisomy 21 (Down syndrome)	512	79.7	50.7	1.2	75.8	62.4	60.0		

 $^{^\}star$ $\,$ Enrollment data base for DHSS (all programs) and DMH (First Steps) NA: Diagnosis not available from birth certificate.

algorithms using child and parent names, date of birth, sex, race, and county of residence plus visual examination of unlinked and multiply linked records.

Data items included in the registry are up to twelve ICD-9-CM-coded diagnoses and program identifier(s), e.g., First Steps and/or CSHCN, with access to data on services provided, family income, and enrollment and discontinuation dates. Program data provide an additional 1.5 percent of the total reported birth defects in the registry (Table A-1).

Pediatric Patient Abstracts. Finally, the registry includes patient abstracts for most inpatient admissions and outpatient visits to Missouri hospitals occurring before the first birthday. Data are collected under the authority and according to the procedures described for newborn patient abstracts. Outpatient records are available for most outpatient encounters at hospitals and ambulatory surgical centers, e.g., outpatient surgeries, major diagnostic procedures, and emergency room (ER) visits. Although abstracts for ER visits not resulting in an inpatient admission are linked to the birth certificate, these data are not included in the birth defects registry because of concern about increasing false-positive cases. Linkage of the pediatric patient abstract to the birth record is accomplished through computer algorithm with limited manual matching of unlinked and multiply linked records. Birth certificates were identified for 95 percent of pediatric patient abstracts. Some unlinked records are believed to represent children who were not Missouri residents at birth. Data items included in the registry are ICD-9-CMcoded diagnoses, ICD-9-CM or CPT-4-coded procedures, type of admission (inpatient or outpatient), admission and discharge dates, hospital identifier, and total hospital charges associated with each hospital admission.

Pediatric patient abstracts provide an additional 27 percent of the total number of birth defects in the registry; i.e., above that from all other components. It is the major source of data on CNS, eye, and digestive system anomalies.

Single v. Multiple Reports. In Table A-2, the percent of defects reported in more than one source is listed. Overall, multiple reports were received for 27 percent of infants with defects, but there is wide variation by type of anomaly. The highest rates of multiple ascertainments were for Down syndrome (80 percent), oral clefts (74 percent), and spina bifida (67 percent). These defects are generally readily apparent and affected infants are likely to require medical attention after the newborn period. Certain anomalies are generally reported on only one source. Over 75 percent of eye anomalies, heart defects, other circulatory and respiratory anomalies, digestive, and musculoskeletal or integument defects were reported only in one data component.

While a high percentage of multiple reports are suggestive of accurate diagnosis, a low percentage does not necessarily indicate a high false-positive rate. Heart anomalies are an example; 56 percent are noted on the newborn abstract and 54 percent on the pediatric abstract, with only 22 percent recorded on any two or more sources. Heart defects noted only on the newborn abstract are likely to be relatively minor defects not requiring follow-up hospitalization, while those noted only on pediatric abstracts are likely to be

those not detected at birth. Pyloric stenosis is an example of a defect that is rarely diagnosed in neonates; pediatric abstracts are almost the only source of data on this defect.

II. Changes in Birth Defect Rates: 1980-2000

Readers who have used data from the birth defect registry for births prior to 1993 will have noticed significantly higher rates in the more recent data. Overall birth defect rates per 10,000 births are listed in Table A-3 by year, using all available data for each year.

In the early 1980s, the overall birth defect rate rose steadily, due to increasing availability of newborn abstracts and more complete recording of diagnoses on those abstracts, but overall birth defect rates for 1984-92 remained in the 330-370 per 10,000 range. For 1996-2000 births overall rates jumped to 510-575 per 10,000. Inclusion of the newly available pediatric abstracts is the primary cause of the increased rates, but increased availability of newborn abstracts and improved ability to link newborn abstracts to birth certificates also contributed to the increase.

Table A-3
Birth Defect Rates per 10,000 by Year of Birth: Missouri 1980-1999

Year	Rate	Year	Rate
1980	260.7	1990	374.3
1981	269.2	1991	374.4
1982	293.8	1992	370.6
1983	309.3	1993	555.3
1984	341.0	1994	540.3
1985	329.5	1995	510.3
1986	363.6	1996	548.8
1987	354.1	1997	562.5
1988	350.6	1998	572.7
1989	364.2	1999	574.5
		2000	572.4

III. Registry Development Schedule

Birth and death data are added to the registry monthly. Reports from other sources are linked and added to the registry at least twice per year. Hospitals submit abstracts quarterly and are allowed five months after the close of the quarter to submit their abstracts. An additional 2-3 months for edits by patient abstract analysts and linkages by registry staff are generally required. Hospital data for out-of-state births to Missouri residents are received only once per calendar year, and a few hospitals fail to submit timely data. Because case ascertainment continues to age one, final data for a calendar

year are available approximately 20 months after the close of the year, e.g., September 2002 for 2000 births. Provisional data sets are used in the interim for projects such as neural tube defect surveillance.

IV. Confidentiality

The Department of Health and Senior Services adheres to strict confidentiality policies and procedures for information included in the birth defect registry and all other departmental data sets. While personal identifiers such as name are needed for record linkage, all data made public are aggregate statistics or are otherwise provided without identifying information. Data are maintained in secured computers, and paper copies with identifying information are stored in locked cabinets when not in use. All DHSS personnel sign oaths of confidentiality, enforceable by state and federal law.

Birth Defect/Disease Category	Birth * Cert. Item	ICD-9 Codes
TOTAL BIRTHS WITH CONGENITAL ANOMALIES	Yes	740-759 excluding 743.65, 744.1, 744.5, 747.5, 750.0, 751.0, 755.0, 756.2 757.3, 757.9, and 747.0 and 752.5 in LBW/preterm **
Central Nervous System Anomalies	Yes	740 - 742
Neural tube defects	Yes	740, 741
Anencephalus	Yes	740
Spina bifida without anencephalus	Yes	741 without 740
Hydrocephalus without spina bifida	Yes	742.3 without 741
Microcephalus	Yes	742.1
Eye Anomalies	Yes	743 excluding 743.65
Anophthalmos, microphthalmos	Yes	743.0-743.1
Congenital cataract and lens anomalies	Yes	743.3
Ear, Face, Neck Anomalies	Yes	744, excluding 744.1, 744.5
Heart Anomalies	Yes	745 - 746
Common truncus	No	745.0
Transposition of great vessels	No	745.1
Tetralogy of Fallot	No	745.2
Ventricular septal defect	No	745.4
Atrial septal defect	No	745.5
Pulmonary valve anomalies	No	746.0
Hypoplastic left heart syndrome	No	746.7
Other Circulatory and Respiratory Anomalies	Yes	747-748, excluding 747.5, 747.0 in LBW/preterm **
Patent ductus arteriosus	No	747.0 excluding LBW/preterm **
Coarctation of aorta	No	747.1
Pulmonary artery anomalies	No	747.3
Agenesis of lung	No	748.5
Oral Clefts	Yes	749
Cleft palate only	No	749.0
Cleft lip with, without cleft palate	No	749.1, 749.2
Digestive System Anomalies	Yes	750-751, excluding 750.0, 751.0
Tracheo-esophageal fistula	Yes	750.3
Small intestine atresia, stenosis	No	751.1
Large intestine, rectum atresia, stenosis	Yes	751.2
Pyloric stenosis	No	750.5

Appendix B continued

	Birth * Cert.	
Birth Defect/Disease Category	Item	ICD-9 Codes
Genital Organ Anomalies	Yes	752 excluding 752.5 in LBW/preterm **
Undescended testicle	No	752.5 excluding LBW/preterm **
Hypospadias and epispadias	No	752.61, 752.62
Urinary Organ Anomalies	Yes	753
Renal agenesis	Yes	753.0
Obstructive urinary defects	No	753.2, 753.6
Musculoskeletal, Integument Anomalies	Yes	754-757 excluding 755.0, 756.2, 757.3, 757.9
Congenital dislocation of hip	No	754.3
Clubfoot	Yes	754.5-754.7
Reduction deformity upper limb	No	755.2
Reduction deformity lower limb	No	755.3
Skull and facial bone anomalies	No	754.0, 756.0
Diaphragm defects	Yes	756.6
Abdominal wall defects	No	756.7
Integument anomalies	No	757 excluding 757.3, 757.9
Chromosomal Anomalies	Yes	758
Trisomy 21 (Down syndrome)	Yes	758.0
Trisomy 13 (Patau syndrome)	No	758.1
Trisomy 18 (Edwards syndrome)	No	758.2
OTHER SELECTED CONDITIONS		
Congenital infections	No	090, 770.0, 771.0-771.2
Congenital pneumonia	No	770.0
Congenital syphilis	No	090
Hereditary anemias	No	282
Sickle cell anemia	No	282.6
Infantile cerebral palsy	No	343
Epilepsy	No	345
Selected endocrine, metabolic diseases	No	243, 270, 271.1, 271.8, 272.7, 275.1, 275.3, 275.4, 277.0, 277.2, 277.5
Intracranial hemorrhage	No	772.1, 772.2

^{*} Congenital anomalies are reported on the birth certificate primarily in the form of check boxes. Anomalies for which a check-box is not provided can be hand-entered and are converted to ICD-9 codes. "Yes" listed for an anomaly category indicates that the category is a birth certificate check-box item, combination of items, or can be obtained through hand-entered diagnosis.

^{**} LBW/Preterm infant: birth weight < 2500 g or gestation < 37 weeks

	Total	Percent		Total	Percent		Total	Percent
County	Births	Linked	County	Births	Linked	County	Births	Linked
 Missouri Total	374,604	94.1	 Grundy	649	94.3	 Pettis	2,796	96.6
Adair	1,376	98.2	Harrison	525	84.2	Phelps	2,269	94.8
Andrew	906	97.4	Henry	1,316	97.3	Pike	996	94.5
Atchison	288	76.6	Hickory	362	93.4	Platte	4,968	94.4
Audrain	1,678	95.6	Holt	234	94.4	Polk	1,745	95.1
Audiain	1,070	23.0	11010	231	21.1	FOIR	1,743	23.1
Barry	2,324	83.2	Howard	5 4 8	98.7	Pulaski	3,069	46.0
Barton	913	94.1	Howell	2,431	86.8	Putnam	303	81.8
Bates	1,025	92.3	Iron	664	98.6	Ralls	452	94.2
Benton	770	97.3	Jackson	49,169	92.3	Randolph	1,647	98.2
Bollinger	681	94.7	Jasper	7,962	97.7	Ray	1,504	96.7
Boone	8,774	97.3	Jefferson	13,716	97.4	Reynolds	356	98.0
Buchanan	5,648	97.0	Johnson	3,340	88.3	Ripley	834	97.5
Butler	2,693	97.2	Knox	268	86.6	St Charles	20,289	97.3
Caldwell	553	97.3	Laclede	2,162	94.5	St Clair	481	96.7
Callaway	2,353	97.5	Lafayette	1,893	96.5	St Francois	3,408	98.7
Camden	1,716	95.9	Lawrence	2,429	92.8	St Louis City	28,043	95.5
Cape Girardeau	4,075	96.9	Lewis	649	75.8	St Louis County	64,354	96.6
Carroll	595	97.8	Lincoln	2,548	97.8	Ste Genevieve	932	98.1
Carter	407	98.3	Linn	884	94.7	Saline	1,413	97.5
Cass	5,523	86.3	Livingston	832	97.1	Schuyler	252	92.9
Cedar	773	95.0	McDonald	1,689	52.8	Scotland	342	65.2
Chariton	392	98.2	Macon	950	94.5	Scott	2,874	97.0
Christian	3,701	91.1	Madison	686	96.9	Shannon	474	92.8
Clark	401	57.4	Maries	506	97.4	Shelby	392	94.6
Clay	12,800	95.9	Marion	2,019	92.8	Stoddard	1,661	97.2
Clinton	1,212	96.9	Mercer	203	62.1	Stone	1,597	87.6
Cole	4,498	98.4	Miller	1,595	97.5	Sullivan	515	96.1
Cooper	920	97.5	Mississippi	1,046	98.5	Taney	2,591	92.5
Crawford	1,559	97.0	Moniteau	987	93.1	Texas	1,318	92.9
Dade	443	93.9	Monroe	530	94.2	Vernon	1,431	94.0
Dade	113	33.3	Monioc	330	71.2	Vernon	1,101	51.0
Dallas	1,027	91.6	Montgomery	702	97.9	Warren	1,570	97.7
Daviess	601	91.7	Morgan	1,097	86.5	Washington	1,553	98.7
De Kalb	542	95.2	New Madrid	1,304	95.8	Wayne	734	97.8
Dent	887	98.0	Newton	3,480	96.4	Webster	2,329	79.3
Douglas	691	93.1	Nodaway	1,044	96.4	Worth	124	98.4
Dunklin	2,407	86.9	Oregon	589	82.7	Wright	1,252	94.0
Franklin	6,384	97.9	Osage	831	97.1	-		
Gasconade	868	97.8	Ozark	515	70.9			
Gentry	402	97.5	Pemiscot	1,830	77.0			
Greene	15,540	91.6	Perry	1,201	98.2			
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